Our IVIG Story: Treating Immune Deficiency
by Jenny Hobbs

IVIG or intravenous immune globulin is something our family first learned about in February, 2005. While my kids have more issues than an immune deficiency and some of this may never be your experience, this is our journey. Everyone is going to have a different story, but there is always enough common ground that we can all learn from each other, especially when something is uncharted territory for your family.

Some children have low levels of the immunoglobulins (or antibodies) IgG or immune systems that do not function well. These children have difficulty fighting infections and are often sick constantly. For these children, IVIG is often an effective treatment. IVIG is a blood product that contains antibodies from over a thousand blood donors to replace a child's missing or nonfunctional immunoglobulins. It is administered through an intravenous line (IV) into the bloodstream approximately every three to four weeks.

Michael's Story

When my son Michael was 15 months old, he had his first hospital stay. It was scary, overwhelming, and a completely foreign experience for me. When your pediatrician sends you from the office to the hospital saying that it would be too long to wait for an
ambulance, it frightens you to the core. That was the first of many hospitalizations for
him in the years to come for infections, metabolic crises, surgeries...you name it.

When he was two, the pulmonologist commented that he was sick a lot more than most
kids his age. We decided not to test his immune system, since there can be a gray area in
whether to treat a borderline immune deficiency. Six months later, when we had been in
the pediatrician’s office at least once a week for 33 straight weeks for ear, sinus, and
throat infections, I asked him if we could proceed with the testing. I still didn’t know
what the options would be for treating, but knew that the situation was getting ridiculous
and it was time to do something.

A week and a half later, we found ourselves at our first immunologist’s office. After
going over his history, she ordered a ton of blood work that would take weeks to get
back. About two weeks later, I got a call from their office saying that his immune system
didn’t respond to his Prevnar or tetanus vaccines and we needed to go in to the
pediatrician’s office to get them repeated. Six weeks later, we repeated all the blood
work again to see if things had improved. The only thing that responded was tetanus, and
his IgG levels, measures of antibodies in the blood, were also low.

Back at the immunologist’s office, we discussed what we would do next. While IVIG
was brought up at this time, the thought of giving my child a blood product every month
was scary, so we opted to try a maintenance antibiotic. Because of his metabolic disease,
a glycogen storage disease that was undiagnosed at the time, his tummy and body didn’t
tolerate the antibiotic and he continued to get sick. Finally, our metabolic team said we
needed to do whatever it took to keep him healthy because his illnesses were having a
profound impact on the rest of his body.

In June 2005, we agreed to consider starting IVIG and sought a second opinion from a
pediatric immunologist, as opposed to the adult immunologist we had been seeing, who
was all that was available where we lived. We made the hour and a half trip to a
children’s hospital where they confirmed that starting IVIG was a good idea at this point.
In August of that year, a week after his third birthday, Michael had his first IVIG
infusion.

Our only option for infusions where we lived was the pediatric inpatient unit of the
hospital. Using a protocol from the immunologist, our pediatrician managed those
admissions, and the hospital staff was wonderful to us. My youngest was nine months
old at this time, so she was with us while my older daughter would play at a friend’s
house on those days. This was our routine every four weeks for the first two years or so.
After about three months, we realized that IV starts were not easy with him and we
elected to have a port-a-cath, a permanent IV or central line placed under the skin,
inserted in December 2005.

**Dealing with Reactions and Complications**
It is possible to get reactions or side effects from IVIG. We realized early on that Michael’s reactions tended to occur about 48 hours after his infusions. He almost always had a headache and vomited about half the time. Since he also has a metabolic disease, these side effects landed us back in the hospital on quite a few occasions. Now he gets pre-infusion doses of Motrin and Benadryl that are then continued for 48 to 72 hours after the infusion as well to try to ward off the migraines.

He also gets his infusions over a longer than usual period, six to eight hours, and we get the same brand every time. Each brand is slightly different, so consistency is key to figuring out which one is best for your child. Keep track of brands, rates, and side effects so you can relay that information to the immunologist and be certain that the brand you are using is truly the best for your situation.

What has helped the most, though, is running a continuous feed through his gastrostomy tube starting the night before his infusion and running for the full 72 hours post-infusion. He has not had to go back into the hospital for IVIG reactions since then. We also keep a stock of Zofran, a medicine that helps nausea and vomiting, to give as needed.

After almost two years of IVIG therapy, Michael started having problems with cellulitis around his gastrostomy site. After three hospitalizations in four months, all of which occurred right before his IVIG was due, the doctors decided to increase his IVIG frequency to every three weeks. IVIG has a 21-day half-life, meaning that after 21 days it is only half as effective. To prevent those fourth week problems, some people will end up needing to infuse every three weeks.

**Meredith's Story**

My youngest, Meredith, started down the same path at about the same age as Michael. In fact, she was hospitalized for the first time on her first birthday. When we started seeing the signs, we decided to test her immune system much earlier. In doing so, we watched her IgG levels dropping when they should have been rising, as usually happens as kids build up their immunity. Since I breastfed her (and Michael) for 14 months, she had my antibodies for a time, but when breastfeeding stopped, she just could not maintain them herself. She, too, was diagnosed with glycogen storage disease only two months after Michael, right before her second birthday.

It was determined just before her third birthday that she would also benefit greatly from IVIG. She, too, had poor pneumococcal vaccine responses and low IgG levels. Because of numerous previous experiences with her and difficult IV starts, we elected to place a port-a-cath before her first infusion. She began IVIG in November 2007, at every three weeks to stay on Michael’s schedule.

**Options for Infusing**
For the next six months, we were saved a hospital room big enough for two beds every three weeks, and the kids infused side-by-side. Meredith didn’t even seem to mind since she had her big brother to follow. We moved in April of 2008 to a completely new state, all new doctors, and a new option for IVIG: the outpatient infusion suite, part of the hematology/oncology center. Since IVIG infusions take longer than most that they deal with at the hematology/oncology center, we were met with some resistance.

We also met a new immunologist that felt maybe IVIG was no longer necessary, mainly due to pressure from the doctors from the hematology/oncology center. Both kids trialed off IVIG in November 2008. During the next three months, they both had strep, sinus infections, and pertussis. Needless to say, we called the old immunologist who ordered immune studies to be drawn. Now we travel every six months to see our metabolic team as well as several other specialists. They agreed we should resume IVIG.

Since we were in a transition phase of leaving the first hospital in the new city, and because it was difficult to find someone to keep our oldest daughter in a new place, we asked if we could start doing IVIG at home with a home health agency. Everyone was agreeable to that, so in February 2009, we had their first home infusion. We are trying to extend the time between infusions back to every four weeks by going up quite a bit on the dosage.

We just completed home infusion number four this past week and we love it! The kids can play in their own environment, sleep in their own beds if they’re tired, and don’t have to be in the hospital. All of our supplies are shipped overnight a few days before the infusion and I just stick the bottles in the fridge until a couple of hours before we plan to start. Accessing their ports (central lines) is still not pleasant, despite numbing cream, but once that is over with, they are just fine. I am the one who accesses their ports, and while I don’t like being the “bad guy,” I know that the same sterile technique is being used each time and that it’s being done properly. I still ask the kids every time if they’d rather one of our nurses do it, but they have been other people’s practice pin cushions before and don’t want that again. Even when I miss on occasion and have to stick again, they still prefer me to be the one to do it.

I have many friends who do subcutaneous immunoglobulin (SCIG or subQ Ig) for their children, which is a less invasive way of administering the same blood product under the skin. One potential side effect of SCIG is cellulitis. Since both kids have had repeated hospitalizations requiring IV antibiotics due to cellulitis, we decided that this was not the best option for our family. We may consider that route down the road, but as long as they are staying healthy on IVIG, I don’t see a reason to switch. SCIG infusions are shorter but done weekly. SCIG uses smaller needles and no IV, but it’s usually two to three needles during each weekly infusion versus our one IV over that same time period. The main advantage of SCIG is that steadier trough levels of the main immunoglobulin, IgG, are maintained.

Our doctors don’t see my kids stopping IVIG anytime soon. Michael has now received it for over half of his life and Meredith is approaching two years. We have made it a
tradition to celebrate IVIG anniversaries. When at the hospital, we would always decorate the room and have cake and other treats to share with the staff. This year, we’ll be celebrating at home. I’m sure our nurses will love to help us celebrate another year of health, thanks to IVIG!