

Information and Support

The Oley Foundation provides its 12,500+ members with critical information on such topics as research, health insurance, and medical advances.

The foundation is also a source of support, helping HPEN patients overcome the unique psycho-social challenges they face, such as their inability to eat, altered body image, and the ups-and-downs of chronic illness.

All Oley programs and services are offered **FREE** of **CHARGE** to HPEN patients and their families.

The Oley Foundation
Help Along the Way

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LifelineLetter

Living with home parenteral and/or enteral nutrition (HPEN)

Drug Shortages Hit Home

Nicole Gerndt, MS, LCPC

I recently discovered the Oley Foundation, which has been helpful to me as I've been learning more about IV drug shortages that have been impacting my son, Finley, as well as others. Finley was born in July 2010. He has been parenteral nutrition (PN) dependent since he was just a couple of days old.

Baby Finley

Finley was born full-term. Until he was born, we had no idea he had a congenital, chronic medical condition. My husband and I were blindsided by it, and had no idea what was in store for the three of us just a little over twenty-four hours after our his birth. It was a whirlwind. To condense a long story a bit, our son, who initially took to breastfeeding like a champ, began to vomit bile and did not pass his first stool (meconium).



Fueled by HPN, Finley loves to run.

He was in for surgery within his first week of life. He was given a high ileostomy and multiple biopsies were taken from his small and large intestine. We were given the tentative diagnosis of hypoganglionosis, which falls under the category of chronic idiopathic pseudo obstruction.

Within the next few weeks of life, Finley underwent a second surgery. They took more biopsies and placed both a central venous catheter and a GJ-tube. They hoped that over time, he would be able to tolerate a slow rate of enteral feedings—even though we had been having little to no success with bolus, NG-, or NJ-tube feedings.

Drug Shortages Hit Home, cont. pg. 2 ➤

Help End Shortages

Mary Patnode

Last year, the Preserving Access to Life-Saving Medications Act was successfully passed through both houses of Congress. This legislation requires manufacturers to notify the Federal Drug Administration (FDA) when and if drug or device shortages might be anticipated. The medical community was assured that FDA-approved drugs and medical devices would again be available for patients/consumers.

This was very good news to the entire Oley community—to consumers, clinicians, and members of industry alike. However, much to everyone's surprise and dismay, shortages of critical drugs have continued, and in some cases increased, in spite of the new legislation. This is a critical problem, especially for home parenteral nutrition (HPN) consumers, who are dependent upon these same drugs for vital nutrients and for whom resources are limited in the best circumstances.

Shortages in HPN

Industries providing HPN solutions in this country are managing in day-to-day, triage mode in order to provide the prescribed and necessary nutrients to their consumers. The shortages are not limited to one



Liz Tucker; Darlene Kelly, MD, PhD, FACP; Mary Patnode; and Harlan Johnson met with Senator Klobuchar's aide Adam Schiff to discuss critical drug shortages.

or two HPN ingredients, but have affected virtually all of the ingredients in HPN. Some drugs are short temporarily or intermittently, while others have been in short supply throughout the crisis. These same shortages have not occurred at all or have occurred with much less frequency and duration in other countries over the last few years.

End Shortages, cont. pg. 3 ➤

Drug Shortages Hit Home, from pg. 1

Going Home

Eventually we were sent home. We struggled to manage everything—hooking our infant son up to PN fifteen hours a day, running GJ-tube feedings at night, changing his ostomy bag multiple times each day, and dealing with

the chronic vomiting that resulted from his inability to tolerate the GJ feedings.

In the early days of GJ feedings, hospitalizations occurred as Finley would become distended and full of bile that was not emptying out of his small intestine. Over time, we learned how to vent his G-tube when needed and use a catheter to help drain stool from his ileostomy. This has helped him tolerate small oral feedings and thankfully avoid an oral aversion, but he could never survive on what little nutrition he takes in orally.

In May of 2011, following a motility study at Children's Hospital of Wisconsin, we were told by our GI team that Finley would always be PN-dependent. Due to his

severe to moderate level of hypoganglionosis (which means he has too few ganglion cells throughout his entire GI system, and the few that he does have are immature/not fully developed) and because of the lack of migrating motor complexes (MMCs), Finley will not be able to rely on oral and/or G-tube feedings to meet his primary nutritional needs. We initially perceived this as devastating news, although it wasn't surprising to us; we had watched Finley struggle with both oral and enteral feedings and absorption issues since birth.

Our Normal

It took some time, but eventually, over the last couple of years, our family has found our "normal" and our routine. Our "normal" involves Finley being connected to his PN for ten hours overnight. The bulk of his nutritional needs are met through the PN. We are now extremely grateful for home PN (or HPN), as without it our son would not be flourishing as he is today. Even more bluntly, he would not be alive. We are also grateful to have had the assistance of a wonderful in-home health nurse who has worked with Finley since he was about six months old.

If you saw Finley in the midst of his usual daily routine, you would see a cheerful, friendly little fella who loves to play in the dirt with his trucks. You'd never know that he has a chronic life-threatening medical condition. We have

been fortunate to have a dedicated GI team that watches his lab work closely and that has made adjustments to his PN as needed.

During our first four weeks in the NICU at the hospital where Finley was born, I was erroneously told by multiple medical professionals that Finley wouldn't be able to live on HPN past a couple of months. I was told he

likely could die due to liver failure and/or while waiting for transplantation. It wasn't until we transferred to another hospital and met with our current GI team that we started to hear that this was not the case, and that many people survive and thrive while on HPN. It has taken me some time to let go of that terrible fear I first felt, and to trust

and believe what I was later told and able to observe, having met others who have survived and done well on HPN.

I am well aware that we will always need to monitor his liver functioning extremely closely in case of detrimental impact to this and other vital organs. We will also always need to be wary and take precautions to prevent central line infections, as these could prove life-threatening.

Calcium and Multivitamin Shortages

Up until recently, we have been able to get what Finley needed in his HPN. Approximately a year ago, we experienced a brief shortage of calcium. Gratefully, this did not last too long, as I am unsure how much Finley is able to absorb enterally, and we needed to give him the calcium through his G-tube.

In the last few months, however, we have been informed that the multivitamin we previously had added each night to his HPN was no longer available. We now have to give a crushed-up multivitamin to him in his G-tube. Again, I worry that he is not getting all of what he needs due to absorption issues. He does not have much intestine, so the vitamins have little chance to be absorbed before they leave his body through his ileostomy.

Phosphorous and Selenium Shortages

A couple of months ago, we were told that phosphorous was no longer available for his



Finley is a cheerful, friendly little fella.

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HPN. Instead we mix a powered phosphorus supplement with two and a half ounces of water twice a day. This may not sound like a lot of fluid, but an extra five ounces a day *is* a lot for a child with GI issues who can, at his best, comfortably tolerate only one to two ounces at a time. Besides causing him physical discomfort—even when we administer it to him in smaller, divided doses—it has caused his ostomy output to increase. This further adds to our worry that he may be flushing out the essential nutrients and supplements without getting much benefit from them.

Finley, who is relatively used to getting medicine through his G-tube and having to take frequent breaks in his play throughout the day due to the need for ostomy care, now has to experience getting more through his G-tube at least three extra times a day. Most times when he knows it's coming, he expresses that it hurts and is bothered by it—which,

as you can imagine, makes for some very fun moments with an active and strong-willed toddler.

While we have not yet had the problems that I hear others have had (needing to rule out renal failure or going to the hospital because of levels dropping too low), within the last couple of months we received feedback from Finley's GI dietitian that his phosphorous numbers were "trending down." I know enough by now to know that "trending down" is generally not good news. Hospitalizations and declining physical health, which can impair his ability to fight off a life-threatening infection, can follow news of numbers and levels "trending down." We know this through prior experience, when his iron or potassium levels dropped too low.

The most recent shortage to affect Finley is selenium. As of May, we have needed to add crushed up selenium to our daily G-tube routine, and we have to coordinate it so it

is not too close to his phosphorous dosage, as it could interfere with the efficacy and absorption.

Future Trends

I am afraid of what IV drug shortages may do to my son's health and physical growth and development should the current shortages continue and if we have additional shortages heading our way.

Having the responsibility and inherent weight of worry caring for a loved one who is dependent on HPN is generally enough, without adding to it the stress of worrying about whether the medication to help them survive will be available and to what extent not having it available may be detrimental to their health.

I want to thank the Oley Foundation for the advocacy efforts and knowledge-sharing they are doing, and would like to offer support by lending our personal experience. ¶

End Shortages, from pg. 1

One pharmacist stated that sterile water was the only ingredient that has not been affected by shortages. This means the HPN supply industry must make decisions about who receives the medicine they need. Who would have guessed that after sweating through the trials of effective medicines, safe techniques, insurance coverage issues, and efficient delivery systems, we would now have to consider availability of needed and sometimes quite common ingredients in this life-saving area of medicine.

"Reach and Teach"

On April 5, Darlene Kelly, MD, PhD, FACP, (recently retired from the Mayo Clinic and now Oley's Science and Medicine Advisor), Elizabeth Tucker (long-term HPN consumer and advocate), Harlan Johnson (my husband, and an advocate and caregiver), and I (a long-term consumer and Oley Board President) met with the Health Care staffer in Senator Amy Klobuchar's office in Minnesota. Oley Executive Director Joan Bishop joined us by phone. Senator Klobuchar had previously

cosponsored one of the Health Care shortage bills and has been an active supporter of quality health care legislation. We spoke with her aide, Adam Schiff, for almost an hour during a prescheduled appointment about our concerns, and we left written material for him to share with Senator Klobuchar.

Our goal was to educate just one of the one hundred senators and to also ask for effective direction to continue our efforts. Adam was receptive to our comments and to our prepared materials. He provided some important recommendations to "reach and teach" as many senators as possible. He said personal stories from consumers are the most effective approach. He also stated that we should target current members of the U.S. Senate Committee on Health, Education, Labor and Pensions, which is the group to decide which bills get attention from the rest of the Senate. Committee members are listed on www.help.senate.gov. or call (800) 776-OLEY for a list.

Personal stories from consumers are the most effective approach....It is our voices that will get their attention!

Speaking Up

I have personally benefited from a regular and reliable HPN industry for almost thirty-three years. While my gut might be artificial and my nutrition may be unique, regular access to needed HPN ingredients allows me to live a healthy and fairly normal lifestyle. I am dependent upon the continued availability of all the ingredients in my individual "recipe" and I will continue to attempt to educate those members of Congress who need to provide leadership and boundaries to the drug companies involved. If our needs cannot be met through legislation, perhaps we need to look for alternative, quality standards. Please consider joining our efforts. It is our voices that will get their attention! ¶

Join us in signing the petition
Oley member Ann Weaver has initiated to urge Congress to end shortages. Link at www.oley.org.

