



PFIC.org Newsletter

Volume 4

Summer 2007

INSIDE THIS ISSUE:

PFIC.org Store	1
Here Comes the Sun	1
Meet Linda, PFIC 2	2
Birthdays	5
Registering on the New Forum	6

PFIC.org Now has an on-line store at: <http://cafepress.com/PFIC>

Last newsletter we introduced two PFIC.org logos to be voted on in the PFIC.org forum. The winning logo is displayed in the upper left corner of this newsletter. Along with the votes came requests for t-shirts with the new logo.

Erin Hovey our Awareness Committee Chairman suggested our group use Cafepress.com to print t-shirts with the PFIC.org logo. Cafepress runs web stores for groups such as ours. They take the orders, print the original art work/logo on t-shirts, coffee mugs, hats and much more. It is wonderful service to small groups like ours that would otherwise have to pay expensive set-up fees in order to offer products with our logo.



Anna in PFIC.org T-shirt

Please check out the web store by using the link <http://cafepress.com/PFIC> or by going to <http://pfic.org> and clicking on the PFIC Store link. For each item purchased \$1.00 to \$2.00 dollars goes to our organization. This money will go to maintaining and improving our web site.

Here comes the Sun

The official start of Summer was June 21st. Did you know that Prograf also called tacrolimus, which is taken by some liver transplant patients, may increase your risk of developing skin cancer? Per the patient information sheet available on Webmd.com, it is recommended to avoid prolonged sun exposure, tanning booths, and sunlamps. Use a sunscreen with a high protection factor and wear protective clothing when outdoors.



<http://www.epa.gov/sunwise/uvindex.html>

The link above will take you to the UV Index Forecast Map. You can put in your zip code or city and get the index value and advisory information for your area.

Meet Linda, the oldest survivor, of PFIC 2

“With good spirit and great faith, you can overcome anything! I am living proof.”

Story by Linda H.

My journey with PFIC 2 began when I was about 4 months old. I started showing signs of liver problems because I was typically irritable, was sleeping poorly and started itching all of the time. I had pick marks all over my skin and there was a tint of jaundice in my eyes. My mother noticed the jaundice right away but the doctors didn't see it. But, my mother was all too familiar with liver problems because her first born, Michael, had them. Michael was born in February of 1961. Within 6 weeks of his birth they knew something was wrong with him. His deep brown complexion and constant fussiness was not normal. An exploratory surgery was performed and my parents were told he did not have any bile ducts. Given the diagnosis of Biliary Atresia, he was given only 6 months to live. Being that it was the early 60's, there was not much else that could be done for him. Much to everyone's amazement, he lived to be nearly 5 years old. They were very hard years for Michael and my parents. With 3 other children under the age of 4 to take care of and Michael needing constant attention, life was not easy. My brother, Bob, was a year younger than Michael and my twin sisters, Sharon & Karen, were 3 years younger. Michael did not grow like a normal child. His stomach was swollen and his skin was very brown. He looked very malnourished and sickly. He had constant nose bleeds, diarrhea and itching. Despite all of this, he was a pretty happy child. He could sit and play by himself for hours. In October of 1965, Michael past away during the night. Because Michael's breathing at night time was very loud and labored, my Mom knew immediately when he had passed away. She was sleeping by his side when his breathing stopped. It was a very heartbreaking night for my parents. Michael was finally at peace but the loss left a hole in everyone's heart that loved him. The actual cause of death on his death certificate says that he died of a very rare lung cancer.



Linda with her husband Darrin sons Michael and David.

I was born on December 4, 1969. When I was almost a year old, I was still fussy and not gaining much weight. My Mom was insistent that the doctor take my bilirubin count. Because of her experience with Michael, she was certain there was something wrong with my liver. Her suspicions were confirmed when my bilirubin came back elevated. The next step was to admit me to Children's Memorial Hospital in Chicago to do further testing. The test they wanted to perform was only available through the military and had very strict guidelines. For three days I was to be strapped to a bed so I could not touch myself in any way and hinder the results. They wanted to collect blood, urine and feces samples. This was a very hard test for us to go through. To have me strapped to the bed like that seemed like some kind of medieval torture to my family. The doctors said I would adjust after a day and I did. It was still hard for everyone to watch. Back then, it wasn't usual to question techniques used by doctors and my parents thought they were doing what was best. The test came back inconclusive and it was back to the drawing board. The doctor told my parents the only way to know for sure if I had bile ducts or if my liver was damaged was to do exploratory surgery because there were no ultrasounds, CT scans or MRI's available back in the early 70's. The surgery was performed before I was released from the hospital. I did have bile ducts and was diagnosed with Dubin-Johnson Syndrome. DJS is a familial conjugated hyperbilirubinemia disorder and has a relatively benign course. A new medication was being released at the time called Questran. Questran was available in powder form and was usually prescribed to people with high cholesterol. It was also prescribed to people to relieve itching associated with gallbladder obstruction. This medication came in a large jar and I was to be given it 3 times a day, mixed in with apple sauce or juice.

For 17 years, I took Questran 3 times a day and my Dubin-Johnson Syndrome seemed like it was under control. Then, in September of 1987, I started experiencing problems. I was loosing weight rapidly and my hands and feet would itch constantly. The Chicago land area experienced a big flood in August and our subdivision lost power for several days and our basement flooded. One of our first thoughts was that I had gotten some kind of virus from the contaminated flood water. I was also going through a lot of changes in my life. I was a senior in High School, my sister had just gotten married and moved out of the house and my boyfriend went away to an out-of-state college. On top of that, I was dieting and not taking my medicine properly. We thought all of these factors might have attributed to my sudden illness. All of the doctors asked me the same questions – could I be pregnant or was I taking drugs or drinking excessively. All of these questions would be reasonable to ask to a healthy 17 year old girl. Although I had started having an occasional drink at parties, with my liver condition I never drank excessively and certainly did not ever take drugs and was not pregnant. My doctor told me because I was becoming a woman, my hormones were changing and it was probably normal that my liver was reacting to it. In December, I turned 18 and was starting to experience more severe symptoms. My bilirubin had skyrocketed to 30 and my skin and eyes were severely jaundiced. The itching was uncontrollable and unbearable. I wasn't sleeping at all because of it and the chronic cough I seemed to get every night. My digestive system was not working at all. Any food I ate would go right through me and I was wasting away to nothing. I had severe nose bleeds that would not stop. I started wearing my glasses instead of my contacts to try and mask my golden yellow eyes but classmates were starting to notice. My strength was weak and I was always fatigued due to the lack of nutrients and sleep. At first, I was dismissed from gym class because I did not have the strength to get through the activities. Right after Christmas, it got to the point that I could not attend school at all anymore. Being halfway through my senior year, I was almost done with high

school. A few required classes that I hadn't finished yet were waved for me through the school district and I was able graduate early. This was a big relief for me as I was getting very self conscience of my looks and I couldn't keep the itching under control.

In February of 1988, the Gastroenterologist we had been seeing performed an ERCP on me to get a better look at my bile ducts. The test was unsuccessful because every time the doctor would inject the dye into my system, I would throw up. With my bilirubin escalating and my weight dropping everyday, the doctor didn't know what to do with me. At my parents' insistence, I was sent to the Mayo Clinic. On a very cold, bitter day at the end of February, my parents drove me on the 6 hour car trip to Rochester, Minnesota. I was put through a battery of tests there including another ERCP. This time, it was successful and it was determined that I had very small, thin bile ducts. A French doctor that saw me thought I should be put on a high regimen of vitamins. He thought the condition came on so suddenly that it might go away just as fast. The other doctors disagreed and thought I needed to be put on a liver transplant list. We were sent home to make a very tough decision and to wait. A couple of weeks after we got home, Dr. McGill from the Mayo Clinic called me with some hopeful news. Through associates of his, he had come to learn about a doctor at the University of Chicago who was treating children with my symptoms by performing a new procedure. How fortunate for us that the hospital was so close to our home! In March of 1988, we went to Wylers Children's Hospital at the University of Chicago to meet with the man who would save my life.

Dr. Peter Whittington was a GI and Hepatology doctor that specialized in a virtually unknown liver condition in children called Byler's Disease. At the time, I believe I was one of the first 20 known case in the world. Basically, my bilirubin was not being excreted properly and it was building up in my bloodstream causing the jaundice and itching. As an alternative to a liver transplant, Dr. Whittington told us that an external biliary diversion could be performed which would include taking a part of my intestine and routing it from my gallbladder to a hole in my lower, right abdominal wall, called an ostomy. Since my bile ducts were so small and thin, the bilirubin would then have another way to be excreted through this alternative route and come out through the ostomy into a small pouch. I would have to wear an ostomy pouch for the rest of my life and the bile that was drained into it would have to be emptied several times a day. He told us we could go home and think about it and let him know. I didn't have to think about it. Given the alternative, a liver transplant, I knew having the biliary diversion was the right decision for me. I was so sick that I was literally on my death bed. What if I didn't get a new liver in time? What if the transplant wasn't successful? The success rate of liver transplants back then weren't as good as they are today. Although the idea of having an ostomy the rest of my life (and at such a young age) did not thrill me, the benefits outweighed the inconvenience of having one. Since I was already 18 years old, I was considered an adult and could sign the consent for surgery myself. Dr. Whittington was puzzled about the fact that I was 18 years old and had lived a fairly healthy life up until then. Since all of the patients he had treated and studied with this disease were children, my case was a bit unusual. How did I get to be almost an adult without any liver complications? Did the Questran really help me during all of those years? Those were some of many questions about my case that may never be answered. Dr. Whittington was sure of a few things though. I did not have Dubin-Johnson Syndrome and my brother, Michael, probably did not have Biliary Atresia. He was certain Michael had Byler's also and tried to obtain his medical records and tissue samples from Children's Memorial Hospital but was unsuccessful. The hospital did not keep medical records from that long ago so we will never really know for sure. Dr. Whittington also diagnosed me with Gilberts Syndrome. Gilberts is a common, benign liver disorder which causes indirect bilirubin to be elevated. Gilberts does not require any treatments and does not cause any complications.

On April 13, 1988, my biliary diversion was performed by Dr. Jean Emond at the University of Chicago. Because Dr. Emond was a liver transplant surgeon, my surgery was postponed several days due to emergency liver transplants. When the time for my surgery came, I was more than ready. Much to our disappointment, I did not get immediately better after the surgery. I did have some relief from the itching and other symptoms I was experiencing, but only about 50%. I also had problems with my ostomy bleeding all of the time. After about my 2nd trip to the ER to control the bleeding, Dr. Emond put another stitch in my ostomy and the problem was solved. Since my quality of life was not as good as it should have been, Dr. Whittington decided to try a new clinical trial drug that was not yet FDA approved. I started taking oral goose bile to see if it would relieve the rest of my itching. It did not so Dr. Whittington decided to try oral bear bile called ursodioxolic acid or Actigall. This medication was successful and I was sent to a children's hospital in Ohio to be put on the clinical trial so I could keep getting the medication. Between my ostomy and the Actigall, my itching was relieved and my body started functioning normally again. I was able to go to my local community college and get a part-time job assisting a child care teacher at the school. In the summer of 1989, I found a temporary, full-time job at a construction office. When the fall came, I was able to keep that job on a full-time basis (32 hours) and go to college full-time. I went to school in the mornings until 11:30am, then went to work from noon-6pm Monday through Thursday. On Friday, I did not have classes so I worked a full, 8 hour day. I needed to do this so I could get health insurance. My body was getting stronger and my life was becoming normal again.

I got married to my husband, Darrin, on August 28, 1993. He accepted me and my ostomy and the possibility that we may never have children due to my liver condition without any qualms. We were young, both 23, when we were wed. My life was very normal. I would go to work all day, come home and exercise, make dinner and spend the evening with my new husband. We would go out on weekends with friends to dinner and to movies or bars. We were very happy. Approaching our 5 year wedding anniversary, we started talking about the possibility of having children. We even went to see Dr. Whittington to ask him what he thought my chances of having children were without causing any liver complications. He told us that he really didn't know because no one with my condition had any children. But, he thought I should go for it. He also told me that Byler's was now called PFIC (Progressive Familial Intrahepatic Cholestasis). They were looking for the gene that causes this condition and asked if me and my siblings would take a blood test for him to do some testing. I said that we would and we went home to ponder our decision. It turned out; we didn't need to make a decision at all. In March of 1998, I found out I was pregnant. We did not plan the pregnancy and even used protection. I took this as a sign from God that the pregnancy was meant to be and that everything would be alright. We were concerned that my bilirubin would escalate during the pregnancy and the itching would return when, in fact, the opposite was true. My bilirubin count was lower than ever. I had no complications, not even regular pregnancy complications like nausea and vomiting or swelling. I felt great during my pregnancy and everyone said I looked better than ever.

Michael was born on October 23, 1998 (named after my brother). I even went to work that day! Although I pushed for over an hour, my labor and delivery was quick. I went into the hospital about 1:30pm and Michael was born at 6:56pm. He was 18 days early and weighed 6 lbs.14 oz. About the last month of my pregnancy, I started getting itchy again. After Michael was born, my bilirubin continued to go up and my itching increased. I started getting jaundiced and having bowel issues again. Dr. Whittington put me on Rifampkin to try and relieve my itching but said it was a temporary fix. I did start to feel better until, one day, my leg hurt so bad that I could not walk. I went to see my family physician who was immediately concerned about a possible blood clot in my leg. He sent me over to the hospital for an ultrasound and blood work. The ultrasound came back normal but the test results showed that my blood was not clotting. The Rifampkin was depleting my vitamin K so I had to get a series of vitamin K shots. I decided to stop taking the Rifampkin since it was only delaying the inevitable. It took about 6 months to start feeling better and a good year for my body to get completely back to normal. I ended up being on an extended pregnancy leave due to my liver complications and went back to work part-time when Michael was 5 months old. After going through all of that, we decided we would only be having one child.

In the summer of 2001, I started thinking about having another child. Michael was going to be three and Darrin was making enough money to support us if I quit working. Darrin was skeptical about this after the first pregnancy but I decided all of the itching and complications were worth it and I didn't want Michael to be an only child. I got pregnant the first month we tried and the pregnancy seemed to be going well until I was into my 15th week. When I went to the bathroom, I saw some very slight bleeding. I immediately had a bad feeling about it. Darrin was out of town for work and I called my OB doctor to tell him I thought I was spotting. He assured me this was normal and not to get worked up about it but, if it would make me feel better, to come into the office the next day for a checkup. At the doctor's office the next day, the baby's heartbeat could not be found so I was sent for an ultrasound. My parents and my sisters were encouraging, telling me everything would be OK but I already knew the baby was gone. The ultrasound confirmed that the baby was only measuring to be about 12 weeks old and I must have miscarried a few weeks prior. This was devastating news for us. A D&C was performed a few days later to clean out my uterus. Extremely sad about the loss but still wanting another child, I got pregnant again a few months later when we were given the go ahead to try again. Again, I got pregnant on the first try and the baby was due on Michael's 4th birthday. This time, I started feeling the effects of my liver disease earlier in the pregnancy. The summer was long, hot and miserable. My itching started in July and my Actigall dose was increased. David was born almost a month early on September 27, 2002 at 7:15pm. Ironically, both Michael and David were both born early, on Friday nights and weighed 6 lbs. 14 oz. I had a fast and furious delivery with both of them and Darrin almost missed both of the births because he was traveling. Crazy coincidence!

Once again, my itching and bowel problems returned. This time, I took the Rifampkin and vitamin K at the same time. I just could not handle taking care of two children and not sleeping during the night, not only because of having a newborn but because of the itching. I felt like I missed the first 6 months of both of my son's lives. They were not enjoyable to me as I even had a hard time holding them for a long length of time due to the itching. I always think how different things would have been if I would have been healthy after their births. How much more enjoyable my time with them would have been. But, I cannot complain. I am very lucky that God has blessed me with two beautiful, healthy boys. I'm so lucky that my liver was strong enough to handle the births. The pregnancy with David did scare me though. My liver and spleen were enlarged and I was terrified that something terrible would happen to me and I would have to leave my two beautiful boys. We would definitely not be having any more children! Dr. Whittington told me that although he would always be there for me, he could no longer treat me. Since he was a pediatric doctor at Children's Memorial Hospital (he left U of C Hospitals), there was only so much he could do for a 33 year old woman. He sent me to see Dr. Richard Green at Northwestern Memorial Hospital. Dr. Green would now oversee all of my liver care and I would get a liver ultrasound and blood work done every 6 months.

It took about a year again to start feeling "normal" again. I was content, and busy, being a stay-at-home mom. With two young boys and a big house to take care of, I had lots to do. Then, the year David was going to turn 3, we decided I needed to find a part-time job. We needed the extra money and I needed to get out of the house a little. David was getting too attached to me. So I got a job at our local library. I wasn't making a lot of money but it was close to home and I made enough to pay our car payment every month. I was only working at the library about a month when a growth was found on my pancreas during a routine liver ultrasound. Dr. Green sent me to a specialist at Northwestern to have a EUS performed (internal ultrasound). A biopsy was taken of the growth and it came back benign. I was told it was only a cyst and would probably never amount to anything serious. We were relieved. We celebrated by going on a dream vacation to Disney World in May of 2006. It was the perfect vacation but, one night, watching the fireworks above Cinderella's Castle, I couldn't help but start to cry. Would this be the last vacation I would ever take with my family? I couldn't shake the feeling that my pancreas problem was not over.

In October 2006, almost a year after the cyst in my pancreas was found, I went to the doctor because I was running a low grade fever and I had some pain in my lower abdominal area. Since I had just had an MRI done at the end of June, I didn't think it would be anything serious. Because of my liver condition, my family physician, Dr. Hubbard, doesn't like to take any chances when I experience unusual symptoms. He sent me for a CT scan and some blood work, including a tumor marker. The blood work came back normal but the CT scan showed that the growth on my pancreas had grown. He arranged for me to have another EUS at Northwestern. The test confirmed that it was not the original cyst that had grown, but it was a new growth. The growth was biopsied and the test came back A-typical or not normal. I was sent to see Dr. Mark Talamonti at Northwestern to talk about the possibility of removing the tumor. Since this was starting to turn out to be more serious than I had thought and I didn't know anything about Dr. Talamonti or the possibility of pancreatic cancer, I contacted Dr. Whittington. Since Dr. Whittington had never heard of him either, he contacted one of his associates and did some research for me. He found out that Dr. Talamonti was one of the top surgeons for pancreatic cancer. He performed an average of 2-3 Whipple procedures a week and he also specialized in liver cancer. I was definitely in good hands. In November of 2006, we went to see Dr. Talamonti and it was confirmed that the best course of action for me would be to have the Whipple procedure done. Since pancreatic cancer is such a serious cancer, there was no room for error. The tumor could be non-cancerous and I might be fine. But, if the tumor was cancerous and not removed, it could be deadly. The only way to know for sure if it was

cancer was to operate and remove it. Pancreatic cancer is not usually found in its early stages because it is hard to diagnose. There are usually no symptoms until it is too late. The Whipple procedure involves removing the head of the pancreas, part of the stomach, the gall bladder and some of the intestines. This was a big deal! I called Dr. Whittington to ask if he thought my liver could handle this type of surgery. He told me that he thought serendipity had played a role in my life and that most people don't get this chance. He thought I had no choice but to handle it. He said if it were him, he would have the surgery. I asked him if he thought the growth could be caused from my liver condition or so many years of taking Actigall. Since PFIC is such a newly discovered disease and I am the oldest known person with it, he had no data to compare it too. This is probably another question that I will never know the answer too. He also told me that the name of my liver condition was once again changed. I now have what's known as PFIC II or BSEP.

On December 5th, 2006, the day after my 37th birthday, I went into surgery to have the Whipple procedure done. A few hours into my surgery, our worst fears were confirmed. It was cancer but the tumor was on the neck and body of my pancreas, not the head. Dr. Talamonti said he could perform the Whipple procedure but the head of my pancreas looked perfectly healthy. On the other hand, the tail of my pancreas looked diseased, like I had pancreatitis. Instead of the Whipple, he recommended doing a distal pancreatectomy and splenectomy. The neck, body and tail of my pancreas were removed along with my spleen. Since part of the stomach or intestines didn't need to be removed, the surgery was less evasive and my recovery was easier. The tumor was removed and it was confirmed that I had stage II pancreatic cancer. The only reason it was stage II instead of stage I was because two of the lymph nodes that were tested came back positive but they were a direct extension of the tumor and had not metastasized (which means it hadn't spread). It wasn't the good news we were expecting but we are optimistic that it was caught in time and I would have a full recovery. I just finished two parts of my treatment. I completed a cycle of Gemzar in January and February. Gemzar is a chemotherapy treatment used in pancreatic cancer. I received a chemo infusion once a week for three weeks. I had a break for one week then I started a 5 ½ week treatment plan of 5-FU chemo and radiation. For the 5-FU, I wore a fanny pack that contained the chemo and it infused into a port in my chest for 24 hours a day, 5 days a week. The radiation was also 5 days a week for 28 treatments. I successfully endured these treatments with very little complications or side effects. I now have a month off from all of my treatments. On May 7th, I start another cycle of Gemzar. I have three more of these cycles to complete before I am done with all of the treatments.

It has been a long haul for us. I have had to overcome a lot of medical complications in my lifetime and I will probably face many more. I am only 37 years old and have overcome two very rare and serious illnesses. If PFIC isn't rare enough, pancreatic cancer in such a young person is possibly even rarer. I have a lot of faith in God and I know he will get me through any challenges I may face. My journey is just beginning and I know God has a great plan for me. He's given me two beautiful boys to raise and I still have a lot of work to accomplish. This new health challenge has made me a better person. There are so many supportive, loving people in my life right now. The unconditional love and support I have received has made me believe more, it's made me love more and it's made me enjoy my life more. When I am done with all of my treatments, I want to focus on helping others with liver problems and cancer. If I can give hope to just one person, it is worth it. With good spirit and great faith, you can overcome anything! I am living proof.

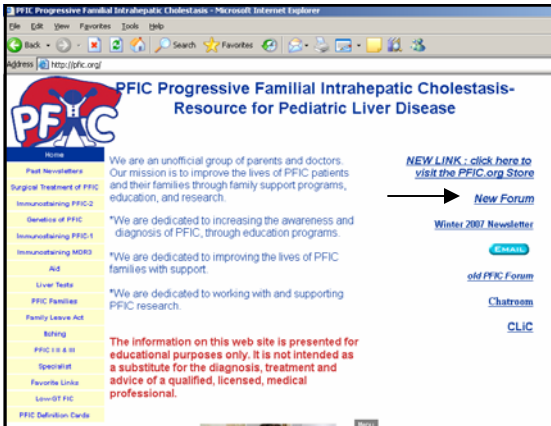
A Special Day to Celebrate

Happy Birthday Zoe!
Zoe turned 10 on July 9th.



The New PFIC.org Forum *Please Re-register

PFIC.org has done a software up-grade. Besides the new on-line store we have new forum software SMF 1.1.3, Simple Machines Forum. Our old forum allowed spammers to register as members. While they could not post on the forum topics, they were able to advertise un-family oriented web pages in our member listing page. We had over 450 spammers advertising on our old forum, with more adding to it every day. The new forum will prevent that from happening.



This new forum will also be more private. Guests to the website will not be able to view posts on the forum. The only drawback to the new forum is that everyone will need to re-register and I will have to re-approve registrations. This measure is to prevent spammers from getting onto the forum. The new forum also has some nice features including a calendar, and the ability to have pictures as part of your profile.

The old forum topics and posting will not be able to be brought over to the new forum. Currently if you click on the 'old forum' you get a message that it is down for maintenance. I will be removing all spam advertising and then bringing the 'old forum' up in a read only mode. All new posts should be made in the 'new forum'.

Please remember to take a minute and re-register on the new forum.

Robin Marceca

PFIC.org Administrator

Instructions for Registering on the New Forum

1. After clicking on New Forum from the PFIC.org home page you should see the long on screen. Choose "Register"
2. Once approved you will get a message in your e-mail and can now click to create your profile.
3. Click on PFIC.org Forum to go to topics and posting.
4. Tabs at the top can be clicked on to view other features such as the calendar

