



PFIC.org Newsletter

Volume 5

Winter 2007

INSIDE THIS ISSUE:

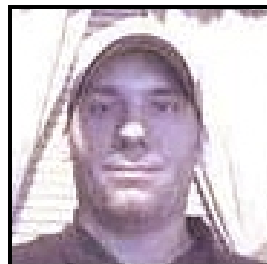
The First PBED	1-3
Member List	3
PFIC Logo	3
Birthdays	3
Thomas's Story	4
Liver Families Quilt	5
E-mail Accounts	6

The First PBED

By Shawn Roebuck

Robin's Note:

Many times when talking about PFIC, I use the words "PFIC kids". Then last month I logged onto PFIC.org and found Shawn's posting. I read it and realized I was going to have to change as "kids" no longer applied. Shawn is a 29 year old PFIC survivor. His story is one that is amazing. When Shawn was diagnosed, PFIC did not have a name, nor did it have a treatment plan. He was not expected to live to be an adult.



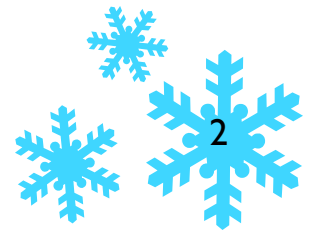
As a child Shawn had the first PBED surgery as a hopeful attempt to treat this un-named disease. When I asked Shawn to write his story he remarked he would but that his life was rather normal. I emailed him back that his normal was awesome. His surgery paved the way for all PBED surgeries, and gives us all hope that our kids too might enjoy a normal life. Here is his story I hope you find it as courageous and inspiring as I do. -Robin Marceca

My name is Shawn Roebuck and I am 29 yrs old. This is my story living with Progressive Familial Intrahepatic Cholestasis. I was born in February 26 1977 in a small town called Vernon Alabama. I have lived in Aberdeen Mississippi most of my life. Between the ages of 5 months to a year I had jaundice and severe itching. I don't remember much, but from what my mother told me I used to itch a lot and dig at my skin. I dug so much they had to put cooking mitts on my hands but that did not help because some how I always got them off.

The doctors where I lived, around Tupelo Mississippi, thought it was just skin allergies but my mother did not believe that. She took me to LeBonheur Children's Hospital to see Dr Peter Whittington and his brother Dr Gene Whittington. They diagnosed me with what is now called PFIC. Without treatment they gave me to the age of 6 to live. They decided to try a new procedure that was called a cholecystostmy tube which drained some of the excess bile salt. It helped relieve some of the itching, but had to be drained a few times a day. I remember a few times when the drain came out and I had to be rushed at the local hospital in Amory Mississippi.



After having the drain for a bit and seeing that it was helping, Dr Peter Whittington and Dr Gene Whittington decided to do a new procedure called cutaneous biliary diversion. At the time I was only three years old and would be first patient to have this surgery done. The procedure is where they took a small piece of the small intestine and hooked it up where it drains the excess bile salt into a colostomy bag, which would need to be emptied at least 8 times a day.



After this procedure my itching stopped and I had no jaundice. It was night and day and we were very thankful. I had followed up with visits with Dr Peter Whittington until he left for Chicago. Then Dr Gene Whittington took over my care. I remember many tests especially the biopsies, because I use to play bingo with my mother as I lay on my side.

I had as much of a normal life after the last surgery as possible, I even played little league baseball. I went to school up to ninth grade. I quit school for un-related reasons and got my GED. The only main problem's I had growing up with PFIC and the colostomy was that a lot of the times my colostomy did leak. I also got really tired quickly and I do not have the strength that I should. But that did not stop me from trying my hardest to live a full and normal life.

At age 15 my mother got me on disability. Everyone in my family told me that I should work or try to work. So, at the age of 18, I got off of disability and decided to work the hardest job I could find, construction jobs. I did everything from shoveling to carrying heavy equipment. My body still tired easily. I would work for 10 minutes and then take a 15 minute break. I did not want to quit the job no matter how tired I felt. I have to thank my mother for giving me the never quit attitude.

I did construction for five years. One day a freak accident happened where I was working on a three story building and the ladder slipped. I fell two stories and crushed my ankle in one hundred pieces. Two surgeries later, I had two plates and twelve screws put in my ankle. I could not run anymore and I could not stay on my ankle much without it hurting. I tried to go back to work but was unable to continue working and made the decision to get back on disability.

Not working was a complete let down for me. I always told myself never quit and never give up, but that day my health and then my ankle told me I could not hold up to it. I am still disability and suffer from depression and anxiety. I live behind my mother in Aberdeen Mississippi. Without my mother, step dad, and family I don't know what I would do. They are my rock and if you get anything out of this story please get this one thing. If you are a parent of a child with liver disease or PFIC you are the rock in the hard times your child will face. You are the support when they have to have there biopsies or blood work and with your love they will get threw it.

This is what I do today. I try to help families with PFIC and other liver diseases cope with the disease and/or the loss of their child. I also spend time with my little sister Chelsey, who is 6 yrs old. I would not change anything in my life. I feel lucky and very grateful, even though my life has not been perfect I am still alive and have a positive outlook on life. If I had one wish it is that every kid and adult with any liver disease, where it be PFIC or other disease would have a full life and a good healthy life.

This is my story and I would not change anything, because the battle with PFIC disease has made me a stronger person.

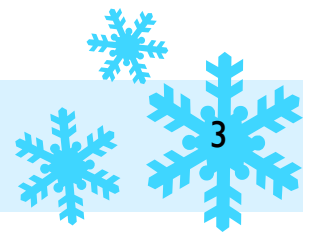
Shawn Roebuck

Shawn@pfic.org

Reference Research Articles below from Shawn's PBED Surgery

1. Partial external diversion of bile for the treatment of intractable pruritus associated with intrahepatic cholestasis. Gastroenterology. 1988; 95 (1) :130-6 (ISSN: 0016-5085)Whittington PF ; Whittington GL
2. Selective Surgical management of Progressive Familial Intrahepatic Cholestasis (Byler 's Disease) by Jean C. Edmond and Peter F Whittington; Journal of Pediatric Surgery V30, No12,

Member Listing Update



3

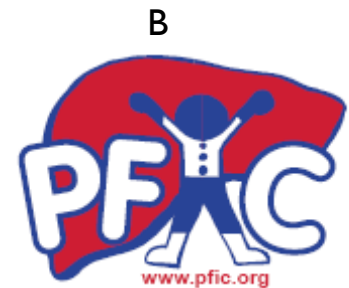
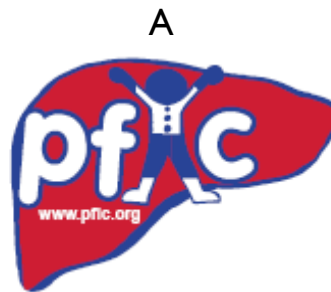
You may receive an e-mail asking for updated information. We are compiling a member listing. The information is for our use only, no information will be given away or sold. Shawn Roebuck is heading up this project. If you would like to be added to the listing or have questions please e-mail him at Shawn@pfic.org

Logo for PFIC.org

Put your choice on the Forum Page

There have been some web-updates including a web page that we can add our logo to. Erin, Cade's Mom and Shannon, Treyton's Mom from LiverFamilies.org came up with these beautiful designs. Obviously Erin and Shannon are very talented.

But, they have left the hard part to us, and that is which one to use. Log on to PFIC.org Forum *Choose your Logo* and choose for Logo A or Logo B.



A Special Day to Celebrate

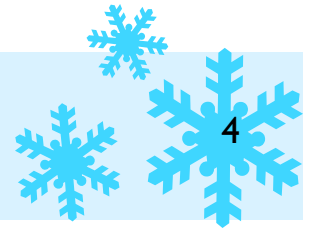
Happy Birthday Cade

Cade will be 3 on April 15.



Thomas's Story

By Mom Amy



Thomas is our 1yr old cutie who was recently diagnosed with PFIC-2. His medical problems began at 4mo when we noticed abnormal bruising on his back. At the time, he was an otherwise happy, healthy baby who was just a lousy sleeper. The bruising brought him to medical attention, and this is when we discovered he had liver problems. He was hospitalized at the time and given a diagnosis of likely EBV hepatitis. However, things did not improve as one would expect and he had a liver biopsy in July 2006. This is the first time we learned of PFIC. Within 2-3 months, he was given the definitive diagnosis of PFIC-2. Currently, Thomas' liver labs and his itching are pretty well controlled with medications. We pray every day for a miracle, but more realistically, that God will ease his itching and for Thomas to keep that amazing smile and personality that wins the heart of everyone he meets! We are so grateful to God for our little Thomas and his sweet, adoring sister Caroline. Thomas is an inspiration to us, for he has been the strongest little guy through all of this and has been very forgiving, especially given all of the testing he has been through.

Liver Families Quilt

From the forum post on Liverfamilies.org

About the Liver Families Quilt

Liver Families Quilt

If you are a LF member, and you have or are related to a child affected by pediatric liver disease; or if you are a young adult who was affected by pediatric liver disease, we would like to invite you to participate in the Liver Families Quilt project. The Liver Families Quilt will be in honor and in memory of the reason why Liver Families exists, and will help spread the word that liver disease and transplant affects many children. In addition to each quilt square, you are invited to write a small story about the meaning of your quilt square, & to include a picture of the child for which the square is being created for.

You do not have to know how to sew to make a square for the quilt.

There is no deadline for this project, and this project will always be ongoing.

The Liver Families Quilt Stories

In addition to your quilt square, you are encouraged to write a story telling us about your square. What made you decide to choose the items you did for the square? Are the colors or fabrics of your square significant? We encourage you to also tell us about your child. Along with your story, you are invited to include one picture of your child which will stay with the story of your quilt square. Your story and child's picture will be included in a binder, "The Liver Families Quilt Stories", which will always stay with the Liver Families Quilt.

The Virtual Liver Families Quilt & Quilt Stories

The Liver Families Quilt Squares, Quilt Stories & Pictures will also be displayed online on the Liver Families website in a special area designated for the Quilt.

How to Participate Quilt Square Specifics:

- 1 quilt square per child affected with liver disease and/or liver transplant will be allowed
- quilt square must be an 8 inch square, with a 1/2 inch space around all sides (keep any design within 7 inches of the square to allow for seaming)

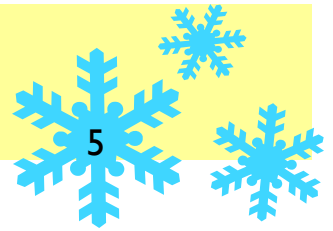
*note: quilt square must be specific to this size, or it may not be able to be added to the quilt & may be returned to you

- do not add backing or quilting to the back of your square...leave it unfinished

Continued-

Liver Families Quilt

From the forum post on Liverfamilies.org



- you do not have to know how to sew to make a square...simply cut an 8 inch square piece of fabric and add momentos if you wish
- you may make your square as simple or as ornate as you desire
- use any fabrics you wish (sentimental things are welcome too... pieces of blanket, sleeper, shirt, etc.)
- you may add any embellishments you wish, but be sure that they are attached securely
- you may add your child's first name, birth date, and death date if applicable (remember the quilt will be displayed online)
- do not use glue on your square
- you may add photos that have been transferred to the fabric
- you may use permanent markers or fabric paint to design your square if you wish
- known copyrighted works (poems, quotes, etc.) that are added to any square must have the source of the work cited directly with the work

Liver Families Quilt Stories Specifics:

- you are encouraged to write a story to share the meaning of your quilt square, about the child it represents, & what this project meant to you
- story must be submitted on an 8 1/2 x 11 inch (standard size) sheet of paper, and must fit on one side of the paper
- you may include 1 unattached picture of the child for whom the square is being created for which still stay with the Quilt Story
- picture can be any size, but no bigger than 5 x 7 inches

Mailing your square and stories

Disclaimer: By submitting your quilt square, story, and/or picture, you are allowing Liver Families to reprint your story, to display the quilt square online, to display the story online, and to display any included pictures online. You understand that the Liver Families Quilt & the Liver Families Quilt Stories and pictures may be displayed and/or shared in public, which includes but is not limited to being displayed on the Liver Families website.

- on a separate sheet of paper, construct and include an information sheet with the following:
 - * the name of the child the square is in honor/memory of
 - * child's date of birth (& date of death if applicable)
 - * your Liver Families member name (your screen name on the website)
 - * your relationship to the child
 - * your full name
 - * your mailing address
 - * your phone number
 - * your email address

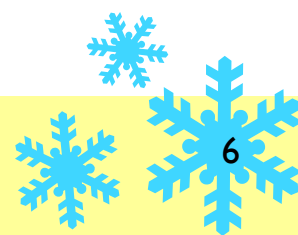
Note: Your personal information included on the information sheet will not be shared outside of Liver Families. The information sheet needs to be included so that we know how to contact you should there be any questions concerning your quilt square or story/picture submissions.

Mail your square, story, picture, and info sheet together to:

Liver Families
23974 Aliso Creek Road, # 242
Laguna Niguel, CA 92677
USA

- Please place your square in a plastic bag to ensure it arrives safely
- Please try to avoid bending or creasing your story, square, or picture and mail accordingly
- It is your responsibility to ensure safe delivery...it is suggested that tracking or delivery confirmation be added when mailing
- **Note:** Your square, picture, & stories will not be returned. It is suggested that you take a picture of your square before you mail it.

Need An E-mail Address?



Did you know as part of our web-hosting package we are given e-mail accounts? If you would like an e-mail account let us know, e-mail PFICII@pfic.org. The account would be yourname@pfic.org

Baylor College of Medicine Medical Genetics Laboratories Has Commercially Available PFIC Test

The CETT Lab mentioned in the Fall Newsletter has made PFIC tests available for the general community. These are the first commercially available tests for PFIC 1, 2, & 3, in the country. CLiC helped make this happen. For more information on testing go to:

The weblink is : <http://www.bcm.edu/geneticlabs/tests/alltests.html>

OR

<http://www.genetests.org>

