

# LiverLink

*The Newsletter of the Alagille Syndrome Alliance*



## **April is..... Organ Donor Awareness Month**

### A Thankful Heart

As we celebrate National Organ Donor Month, I am brought to a point of deep gratitude. For me and my family, Organ Donation and Transplantation have taken on a deep meaning in the past three to four years. We have twin sons with AGS and one of the twins, Timothy, had very poor liver function. Function so poor that the only resolution was a Liver Transplant. For the first five years of his life we saw his activity level and health decline dramatically until that day that the phone rang to advise us that a donor organ was procured and Timothy was to be the recipient. We know very little of the donor other than it was a young child and they lived somewhere relatively close to Cincinnati. While we have sent (through the hospital) a thank you, we do not know who this family is. We are forever indebted to them as it could not have been easy to make such a decision while grieving the loss of their child. It is our hope that somehow and in some way they can know that their decision has forever changed the life of Timothy and the entire Caldwell household. Life has never been the same. The child that had little desire to even walk because of the xanthomas on his feet, now runs constantly and is pursuing things that looked impossible three years ago and all of this was made possible by the decision of one family to make a difference through organ donation. May we all be willing to make that decision should we be placed in the same situation.

In sincere Gratitude to that family that gave so much and all of those like them who have and are registered to be organ donors.....

Thank You

Richard Caldwell for Beth, Zac, Chris, Tim, and Matthew

#### **Inside this issue:**

PRESIDENTS PAGE	2
DR. KATHY LOOMES	3
IN THEIR OWN WORDS.....	4 &5
AGS PICNIC	6
Rx PATIENT ASSISTANCE	7

LiverLink is published six times per year for members of the Alagille Syndrome Alliance, a national support network for people with Alagille Syndrome (AGS), a rare inherited liver disorder. The primary purpose of LiverLink is to provide general information. LiverLink does not provide medical advice, nor does it promote, endorse, or recommend any product, therapy, or institution. Its contents should not be used for diagnosing or treating health disorders. Readers are advised to seek advice from licensed health professionals regarding AGS or other disorders. Statements and opinions expressed in articles are not necessarily those of the Alliance.

Contributions to LiverLink should be sent to: Cindy Hahn, President, Alagille Syndrome Alliance, 10630 S.W. Garden Park Place, Tigard, OR 97223. 503-639-6217. No faxes please. Copyright©2002 Alagille Syndrome Alliance. All rights reserved.

## President's Page

Last issue I waxed poetic with my friend Robert Frost. This time I've hunkered down to share some highlights of our recent annual meeting of the Alliance Board of Directors. It was an action packed two days, one of which included a 12-hour marathon meeting, focused on the Alliance and the wonderful AGS families we serve. Inspiring, energizing, motivating, and yes, exhausting, aptly describe it. Here are some highlights:

- We've added awareness and research to our purpose statement, so it now reads:

***The purpose of the Alliance is to serve as the main networking resource and source of information for people with AGS, their families, friends, and health care providers. The Alliance also is dedicated to increasing public awareness of AGS and supporting research efforts on behalf of the AGS community.***

- We've also added research as a sixth goal, so our goals now include:
  - Support Network: To provide a support network for people whose lives have been touched directly or indirectly by AGS.*
  - Family Gatherings: To provide a forum which enables AGS families to meet one another on a regular basis for mutual support and sharing of current information on AGS.*
  - Gather and Distribute Information: To gather and distribute information about AGS, research studies, and helpful services to AGS families, friends, and health care providers.*
  - Increase Public Awareness: To increase public awareness of AGS.*
  - Involve Members in Programs and Services: To involve as many members as possible in Alliance programs and services.*
  - Research: To support research efforts on behalf of the AGS community.*
- We've chosen Atlanta, GA, for our next Symposium – AGS2005 – and tentatively settled on June 23-26, 2005, as the date. Mark your calendars!!
- We're dedicated to publishing *LiverLink* six times a year and, so far, we're on track. The January-February issue should already have arrived in your mailbox, and this issue will join it shortly.
- We're reconstituting our Scientific Advisory Board so its members will more accurately address the many areas of concern for AGS families and be more readily available for consultations and information.

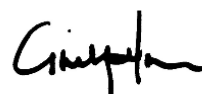
- and -

- We're setting our sights on 2007 to host the first ever AGS Kids Summer Camp. Stay tuned for more info...

There are numerous other objectives for programs and services we plan to continue or offer for the first time, and we are still searching for a “celebrity spokesperson” to represent the interests of the AGS community. But the above list gives you a taste of our Board's productivity and enthusiasm for everything AGS. Rest assured that you have a terrific group of representatives working to support AGS families and raise awareness of this disorder and its effects on our special children and adults. I wish each and every one of you could have joined us for this meeting – awesome, electric, with a good dose of humor and camaraderie – you would have been thrilled, as was I!

Look for a lot of new features in *LiverLink*, a snazzy new look to our Web site, and some community-building regional events in the months to come.

With Love and in Peace,



# And the winner is.....

## American Liver Foundation Researcher of the Year

Dr. Kathleen M. Loomes, a Pediatric Gastroenterologist and Hepatologist at The Children’s Hospital of Philadelphia has been named as the Delaware Valley Chapter American Liver Foundation Researcher of the Year for 2003.

Dr. Loomes completed her residency at in Pediatric Gastroenterology and Nutri-phia. During that fellowship, she won the Hospital of Philadelphia. She entered and Dr. Nancy Spinner where she began the development of the liver and in the has been a very active member of the Group at The Children’s Hospital of sively on Alagille syndrome, including at Alagille syndrome sponsored by the Ala-



Johns Hopkins University and her fellowship tion at The Children’s Hospital of Philadel-Fellow of the Year Award at The Children’s into the laboratories of Dr. Rebecca Oakey to investigate the role of *Jagged and Notch* in function of bile duct cells and liver cells. She clinical and research Alagille Syndrome Philadelphia. She also has lectured exten-the regional and international symposia on gille Syndrome Alliance.

We are extremely proud of Dr. Loomes and her accomplishments to date. We antici-pate further successes and appreciate all of her efforts towards understanding the basic problems of Alagille syndrome in the liver. She will be honored at the annual American Liver Foundation gala and ball to be held in Philadelphia on April 4, 2003.



## *From the Desk of.....Dr. David Piccoli*

### Clinical Research and HIPAA

A federal law was passed in 2002 which was designed to protect healthcare coverage, portability and health informa-tion privacy. This Act, abbreviated HIPAA, is the Health Insurance Portability and Accountability Act. Many hospi-tals have been struggling to meet the guidelines and regulations which HIPAA will require to be in place in April, 2003. There are many valuable and important aspects of this Act which should benefit patients and their families.

One of the important safeguards of this Act is to protect private patient information so that it cannot be misused in any way. You will probably notice many changes in hospitals and hospital-associated office practices in the new next few months. The law also has very significant implications for clinical research and patient-related databases. Much of the clinical research into Alagille syndrome has been performed by collecting information from patients and physicians for analysis and study. Although final guidelines at most institutions have not been completed, it appears clear that patient databases for clinical research will have to be totally changed to meet the specific regulations in the HIPAA law. Over the next few months, patients and their families will have to be contacted and give specific permission to have clinical information entered into these databases. Regardless of this permission, patients cannot be identified specifically in the databases and of course will never be identified in any research or publications about the disease.

As we understand more about this Act and its implications, we will be asking families to give permission for their medical information to be reviewed. Without this permission, advances in the care of patients with Alagille syndrome will be severely hampered. Undoubtedly more will follow about this Act in the next few months

# LIVER TRANS-

## DARREN GEHMAN

1. How old were you when you had your transplant? I was nine.

2. What was it like before you had your transplant? I was yellow and itchy, I was tired all the time. I had bumps on my knuckles, and I took a lot of pills.

3. How was the itching before transplant? I had scabs on my arms.

4. Where did you have surgery? At CHOP

5. How long were you on the list? 8 months

6. How long were you in the school? I was in the hospital for of the school year...I think there



hospital, and did it affect two weeks, but it was at the end were only three weeks left.

7. What was the worst part of the oxygen...I woke up and they

transplant? The tube for the wouldn't remove it.

8. What was the best part of the

transplant? The child playroom.

9. How was the itching after sur-

gery? It was just gone.

10. How long before you felt really well after transplant? About a year.....I caught a lot of colds

11. Do you have any advice for other kids who may have a liver transplant? Try to sleep a lot the first night so they take the air tube out, Make sure you go to the playroom and play jokes on the doctor! (In Darren's case, Dr. Piccoli!)

12. How old are you now? 13

13. What are your interests now that you feel better? Bouncing on the trampoline, swimming, playing the game tube and the computer.

14. Can you play sports? Yes, but I have to be a little careful

# IN THEIR OWN WORDS.....

## TIMMY CALDWELL



1. How old were you when you had your liver transplant? I was five.
  2. What was it like before transplant? I scratched till I hurt, and I was tired all the time.
  3. What was it like after transplant? I didn't itch.
  4. How long before you felt better.....more like playing? I felt good when I got home, but Mom says it took a year for me to "catch-up."
  5. How did it affect school? I'm homeschooled. I learned my numbers and how to write my name at Ronald McDonald House.
  6. How long were you on the list? Six months
  7. How bad was the itching before your liver transplant? Bad.....itchies made Disney World bad. (Tim's family had a family vacation to Disney just before transplant.....the Florida heat made Timmy miserable)
  8. Where was your surgery? Cincinnati
  9. How long were you in the hospital? One week. I liked Ronald McDonald House. I had to wear a mask.
  10. How old are you now? 8
  11. What do you like to do today? Play Star Wars, Legos, ships, play outside, play catch, play football.
  12. Do you have any advice for any other kid who may have a transplant? Don't be afraid of the owies...even if you cry, they are good for you.
-

# Annual

## Alagille's



## Picnic

**Attention:** This is not just another annoying piece of junk mail! Don't even think of throwing it in the trash! This is your ticket to a successful summer! Just tear off the bottom and present at the door of the annual Alagille's picnic to be eligible for a complete soaking by a legion of water gun toting kids and kid wannabes. But that's not all! You will also be treated to the latest scoop as to what is happening in Alagille's research and what is new in treating and dealing with the problems associated with the syndrome. As always, the picnic will feature members of the staff from Children's Hospital of Philadelphia who will be presenting information as well as answering your questions. So put a big red circle around July 26<sup>th</sup> and begin making plans to attend. Your summer will not be complete if you miss this event! More details will be coming later.

### Admit One Family

This ticket entitles the bearers to subject themselves to the water fights, feast on the delicious food, and participate in the discussion at the annual Alagille's picnic.

Where: Lowell & Teena Gehman's  
1840 Oaklynn Dr.  
Green Lane, Pa. 18054  
lgehman@excite.com

When: July 26, 2003

---

## Letterbox

**Janice Kruger** writes that she and her daughter, **Emily**, who is eight years old both have AGS. Emily was diagnosed at 10 weeks of age due to jaundice, failure to thrive, and abnormal liver functions. When Emily was one year old, her family was told that she would need a liver transplant when she was eight or nine years old.

By the time Emily was four years old, the Krugers decided to change their lifestyle, sell all their possessions, and travel around Queensland and the Northern Territory of Australia. As Janice puts it: “We escaped doctors, medications, putting Emily into school, and the world of Alagille.”

Just recently, they settled in Katherine Northern Territory. Janice says that Emily has begun asking about her itchiness and would like to associate with other AGS kids. Emily is now a healthy eight years old and takes no medication, although she continues to itch. She recently started school – her own decision – and she hasn’t looked back since.

Janice and Emily would like to hear from other AGS families, to share day-to-day concerns and just to “get to know another AGS family.” You can email Janice and Emily at [kruger.nt@bigpond.com](mailto:kruger.nt@bigpond.com). They’d love to hear from you!



## Rx Patient Assistance Programs on the Web

**RxHope.com** (<http://www.rxhope.com>) is the only patient assistance Internet initiative financially supported by the Pharmaceutical and Research Manufacturers of America (PhRMA) and participating pharmaceutical companies. RxHope.com began as a grassroots effort of the Patient Assistance Managers and Directors of PhRMA-member companies and has grown into the leading Internet-based patient assistance and sampling web portal in the pharmaceutical industry.

**HelpingPatients.org** (<http://www.helpingpatients.org/about>) is a new interactive Web site that provides a comprehensive one-stop link to thousands of medicines offered through hundreds of patient assistance programs sponsored by PhRMA-member companies, non-member companies, and government and local organizations. In 2002, programs supported by PhRMA members helped 5.5 million underinsured or uninsured patients to obtain more than 14 million prescriptions, up from 2.4 million patients and 8 million prescriptions in 2000.

---

Alagille Syndrome Alliance  
10630 SW Garden Park Place  
Tigard, Oregon 97223  
Phone: 503-639-6217  
Email: [alagille@earthlink.net](mailto:alagille@earthlink.net)

---

WE'RE ON THE WEB  
[WWW.ALAGILLE.ORG](http://WWW.ALAGILLE.ORG)

---

## Practical Fundraising for Research and Education

At the Board of Director's meeting in Atlanta, one large point of discussion was that of fundraising. While it may seem like something that big groups do it is actually something that each and everyone of us can help with. Back in January, a letter went out to the entire mailing list giving some very practical ideas for things that we can all do. Each month we will be spotlighting some simple ways in which YOU can help the Alliance continue to meet the needs of the AGS community as well as expand our reach. This month's Tip:

**GARAGE SALE.** Yes it is that time of year when many of us are going through our stuff. A garage sale is a very basic and simple way to generate money for the Alliance and you can partner with others in your communities to make it an event. You may find yourself saying- "But I can't raise thousands of dollars with a garage sale". The reality is that you never know and even if you don't EVERY dollar helps support YOUR support group. Additionally, if a group in New York raises 200 dollars and a group in New Mexico raises 50 dollars and so on and so on-it all adds up quickly.

As always we appreciate ALL of the support you give to the AGS Alliance for the AGS Community.

See ya next LL

## *Alliance Board of Directors*



*We're not sitting around twiddling our fingers and toes... We're working hard for you and the AGS Community.*

**Cindy L. Hahn**, President and Secretary  
10630 S.W. Garden Park Place  
Tigard, OR 97223  
(503) 639-6217  
Email: [cchahn@worldnet.att.net](mailto:cchahn@worldnet.att.net)

**Chris Hahn**  
10630 S.W. Garden Park Place  
Tigard, OR 97223  
(503) 639-6217  
Email: [cchahn@worldnet.att.net](mailto:cchahn@worldnet.att.net)

**Erik Luxhoj**  
3016 E. Lourdes Drive  
Appleton, WI 54915  
(920) 830-6611  
Email: [eluxhoj@new.rr.com](mailto:eluxhoj@new.rr.com)

**Joe Anderson**  
375 S. Depew Street, Apt #103  
Lakewood, CO 80226  
(303) 935-1713  
Email: [joemidget@aol.com](mailto:joemidget@aol.com)

**Beth Caldwell**  
994 Amherst Drive  
Marion, OH 43302  
(740) 382-6948  
Email: [recaldwe@gte.net](mailto:recaldwe@gte.net)

**Rich Caldwell**  
994 Amherst Drive  
Marion, OH 43302  
(740) 382-6948  
Email: [recaldwe@gte.net](mailto:recaldwe@gte.net)

**Patti Everett**  
550 West Magnolia Circle  
Alpharetta, GA 30005  
(770) 740-1910  
Email: [patti-everett@mindspring.com](mailto:patti-everett@mindspring.com)

**Brenda W. Garza**  
380 Southmoor Circle  
Stockbridge, GA 30281  
(770) 954-9504  
Email: [bgarza@cdc.gov](mailto:bgarza@cdc.gov)