

# LiverLink

*The Newsletter of the Alagille Syndrome Alliance*



This issue of *LiverLink* is the third in a three-part series on itching.

If you still have questions, check out the Alliance Bulletin Board at [www.alagille.org](http://www.alagille.org)—families are posting questions about itching and a bunch of other topics!

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## Itching and Alagille Syndrome..... by Ronald J. Sokol, MD

Itching (or pruritus) is a common and very bothersome problem in children and adults with cholestatic liver diseases (liver disorders with poor bile flow from the liver to the intestine), especially those with Alagille Syndrome.

The severity of itching is usually related to how poor the flow of bile is, thus children with more jaundice and higher serum cholesterol levels are usually the ones who have the most severe itching. Itching may be particularly severe in children with very high serum cholesterol levels and fat deposits (xanthomas) in the skin

at the elbows, knees, ears and other sites.

Itching is usually first noticed when the infant reaches about 6-9 months of age and can scratch, although the infant may have been irritable from itching prior to this time. Itching may be particularly bad at night (for unknown reasons) and may interrupt sleep or cause children to scratch their ears, face, palms, soles, arms, and legs to the point of bleeding. Occasionally the skin becomes infected at the sites of scratching, leading to more itching, and requiring antibiotic therapy.

The cause of itching during cholestatic liver diseases is not known. It was once thought that bile salts accumulate in the skin in cholestasis and irritate skin itch receptors. This has now been shown not to be the case. It is now felt that there is something that circulates in the blood in cholestasis that stimulates certain areas of the brain that give people the sensation of itching.

Continued on Page 4

## Bulletin Board—Up and Running!

The Alliance Bulletin Board (BB) is off to a great start! Logon to the Alliance website at [www.alagille.org](http://www.alagille.org) where you'll find instructions on accessing the BB. Here is a sampling of the topics that have been posted so far:

- What are your thoughts about a week-long AGS Kids Summer Camp?
- Who plans to attend Symposium 2002 and where will

you be coming from?

- Are there kids out there who are interested in an AGS Kids Pen Pal Network?
- Are you an adult with AGS? If so, Joe Anderson would love to hear from you!

Next time you're surfing, logon to the Alliance BB and post a new message, or respond to one that's already there. We're thrilled to have this new feature available to

our families and others interested in AGS!



## President's Page

Welcome to Fall! To those of you with school-age children, that means getting swept up into the whirlwind of school activities that tend to dominate the family calendar from now until next June. Our family is no exception! Our twins started Kindergarten this year, in two separate classes so they have the freedom to develop their own personalities and friendships, and Alaina is now a 5<sup>th</sup> grader. All three are enrolled in gymnastics classes, and Alaina is a Junior Girl Scout and member of a Destination ImagiNation team. I've become the family chauffeur and continue to volunteer in all their classes—a role I take very seriously.

Speaking of school involvement, we moved all our children to a new elementary school this year, primarily to give Alaina a fresh start at making friends and an opportunity to avoid the teasing that had become very bothersome at her previous school. I'm happy to report that she's made two really good friends at her new school, is in a new Girl Scout troop that has accepted her for who she is, and has not experienced a single event of teasing since school started.

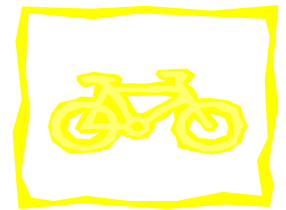
As Alaina's mom, I've found that I have to be a strong and persistent advocate for her in school, explaining

her AGS to her teacher, the principal, school counselor, and P.E. teacher, as well as her classmates—the latter being the most important in the fight against teasing. Alaina asked me to talk to her class about AGS so that they would understand her itching, hives, and difficulty with sitting still, among other things. I visited her classroom on the first Friday of the school year and am so glad that I did—I wish that I'd done this every year since she started Kindergarten! The kids had lots of questions and Alaina and I answered them as straightforwardly as we could. She felt 100% better about having AGS and just being herself with her classmates, and I felt elated that they were so receptive. It was a boost to Alaina's self-esteem and I encourage every AGS parent to do the same in their child's classroom. If you'd like some suggestions on what to say, or a copy of my brief presentation, just let me know.



Before closing, just a brief note about the September 11, 2001, terrorist attacks on New York City and Washington, D.C. The Alliance has numerous families in this part of the Eastern U.S. and it is our hope that all of you are safe and well. It's one thing to watch the events, and their repercussions, unfold from a distance, as I have done over the past weeks. It's entirely another matter to be there and be directly affected by the horrific loss of life and destruction that occurred.

The magnitude of the devastation didn't hit home with me until I received a phone call from Karen Wertheimer at the American Liver Foundation on September 13—she was calling from her home because the ALF office is just three blocks from the World Trade Center in New York City and they were trying, as best they could, to continue functioning in the midst of utter chaos. She informed me that everyone with the ALF was safe and accounted for; she was working from home because she felt that she needed to do something to feel "normal" and to take her mind away from the disaster that had struck just two days prior. It is at times like these, even more than in "normal" circumstances, that we must all pull together to support and look out for one another, and to cherish one another and the community to which we belong.



**"As Alaina's mom, I've found that I have to be a strong and persistent advocate for her in school, explaining her AGS to her teacher, the principal, school counselor, and P.E. teacher, as well as her classmates—the latter being the most important in the fight against teasing."**

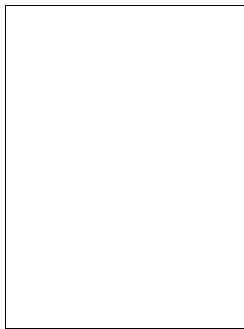
Until next time, with hope and in peace,

Cindy L. Hahn, President

*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©2001 Alagille Syndrome Alliance. All rights reserved.

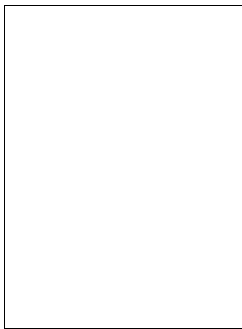
## AGS for Kids: A Story for Alaina’s Classmates... by her Mom, Cindy Hahn



“Alagille Syndrome—it’s a liver disorder, which means Alaina’s liver doesn’t work right. She was born with it, but we don’t know why it picked her and not someone else.”



“Does anyone know what jobs your liver has to do in your body? Well here is a list of just a few of its 5,000 jobs!”



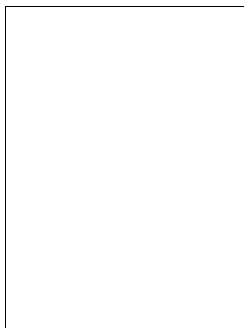
“Alagille Syndrome, or AGS for short, isn’t just a liver disorder. It also affects a lot of other parts of the body. This picture shows those parts.”



“One of the worst effects of AGS is itching. Sometimes Alaina itches so much, she feels like a cat gone crazy! She even scratches in her sleep!”

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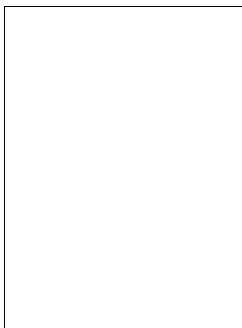
“Alaina asked me to talk to her class about AGS so that they would understand her itching, hives, and difficulty with sitting still, among other things. I visited her classroom on the first Friday of the school year and am so glad that I did—I wish that I’d done this every year since she started Kindergarten!”



“Sometimes Alaina feels like an alien, because she is different from other kids and she rubs her nose a lot and gets hives when she has a cold.”



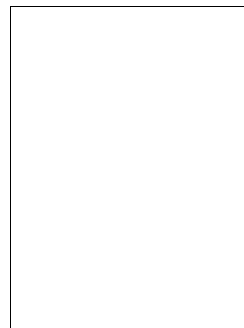
“Alaina gets frustrated and angry about being different, especially when other kids make fun of her and when she feels left out.”



“Sometimes Alaina feels like she has a target painted on her shirt that tells kids, ‘Go ahead and tease me.’ She feels like kids don’t listen when she tells them she can’t help being different.”



“All Alaina wants to be is a normal kid, like everyone else. She wants other kids to like her and to accept her as she is, even if she has AGS.”



“Does anyone have questions? Alaina and I would be glad to answer them as best we can!”

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### Remember...

**the Alliance** on special occasions for loved ones with AGS – birthdays, graduations, transplant anniversaries, or just because... With your help, the Alliance will continue to grow!

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## Itching..... (continued)

These brain pathways may involve the opioid system that also mediates pain relief. More work in this area will hopefully lead to the development of new treatments.

Treatment of pruritus involves local skin care and medications. Keeping fingernails short, covering the hands with cotton socks while the child takes naps or sleeps at night, wearing long-sleeved night shirts to cover the arms, humidifying the air in the child's bedroom at night, applying lubricating creams to the skin after baths, and adding oil to the bath water may help reduce the itching and protect the skin from further damage (and itching) from scratching.

If these measures are not effective, there are several medicines that can be used. Oral treatment with ursodeoxycholic acid (15-25 mg/kg/day) may increase bile flow and decrease itching, although children may have transient increased itching at first for a few weeks. Cholestyramine is a gritty powder that can be taken by mouth 2 or 3 times a day and it will bind bile acids in the intestine and may help increase bile flow and reduce itching. Its side effects are constipation and acidosis, and it will bind other medicines and vitamins that are given at the same time. Therefore, no other medicines or vitamins should be given during the 1-2 hours before or after a dose of cholestyramine. Rifampin is an antibiotic that reduces itching very well, although it may cause liver injury (rarely) itself. It is given twice a day at a total dose of about 10 mg/kg/day. It is not

known how rifampin works, but liver blood tests and a complete blood count should be monitored periodically during its use. We have found rifampin to be very helpful for itching caused by cholestasis.

Other treatments for very bad itching include a partial biliary diversion. This is constructed by a surgical operation that interposes a short loop of intestine between the gall bladder and the skin of the abdomen, with a stoma put on the skin that allows bile from the gall bladder to exit through the stoma into a bag, that is emptied several times a day. In this way, some of the bad chemicals in bile that produce itching are removed from the body and thrown away. The bag is left on indefinitely, because the itching will recur just as badly if the surgical procedure is reversed. This procedure has been done in several Alagille Syndrome patients, with some definite success. Enough patients have not had it done to be able to confidently recommend this procedure, but it should be considered for very bad itching that does not respond to the above treatments.

In other patients, severe, unremitting itching (particularly if other bad complications of liver disease are present) may lead to the recommendation for a liver transplantation which will cure the itching if all goes well.

Finally, research is being performed to determine if drugs that block the opioid receptors in the

brain can safely be given long-term to children and if they are effective for itching. Occasional patients have responded to treatment with UV light therapy administered by a dermatologist.

*Additional information can be found in the chapter, "Medical and nutritional management of cholestasis" in the book, "Liver disease in children", edited by Drs. Frederick Suchy, Ronald Sokol and William Balistreri; Lippincott, Williams & Wilkins, 2001; pages 202-207.*

*Ronald J. Sokol, MD, is an Associate Professor of Pediatrics and Medical Director at the University of Colorado Health Sciences Center and The Children's Hospital, Denver, Colorado. Dr. Sokol is on the Alliance Scientific Advisory Board and chairs the American Liver Foundation's Children's Liver Council, which recently formulated the ALF's Pediatric Liver Research Agenda. The Alliance would like to thank Dr. Sokol for this helpful information on itching and its possible treatments.*



## AGS Angels

**Matthew David Newman**  
August 5, 1989 – September 3, 2001

Matt fought the hard fight with AGS, especially over the past three years. He was listed to receive a liver and kidney transplant and was undergoing surgery to insert a kidney catheter for dialysis when he passed away on September 3, 2001.

The Newmans were one of the first families to join the Alliance—about 6 months after the group started. Matt’s mom Beth would send updates on his condition along with pictures as Matt grew, so I felt like I knew him well although I’d never met him in person. I cried along with Beth when she called me with the news on the night of September 3<sup>rd</sup>.

Several families have sent donations to the Alliance in Matt’s memory, for which we are grateful. I take solace in the thought that Matt is now at peace and free of pain. He shone for 12 years here on Earth, brightly and strongly, and now he shines on us from above. May he be at peace. *–Cindy Hahn, Alliance President*

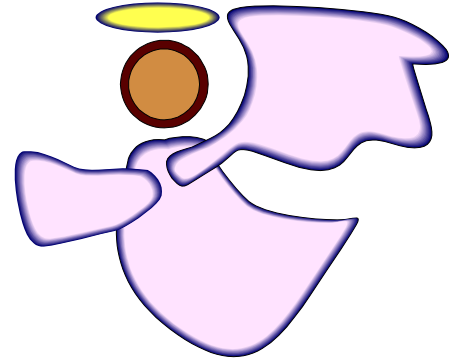
**Brian Clifford**  
May 9, 1988 – May 7, 1999

Debra and Dan Clifford recently sent a donation to the Alliance in memory of their son Brian, who passed away on May 7, 1999, just two days before his 11<sup>th</sup> birthday.

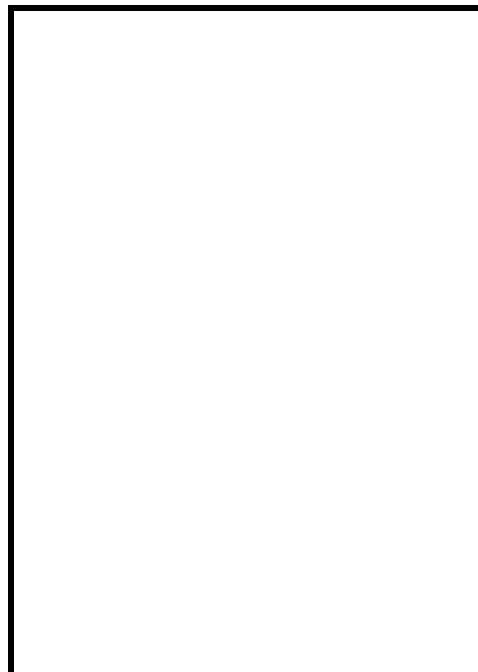
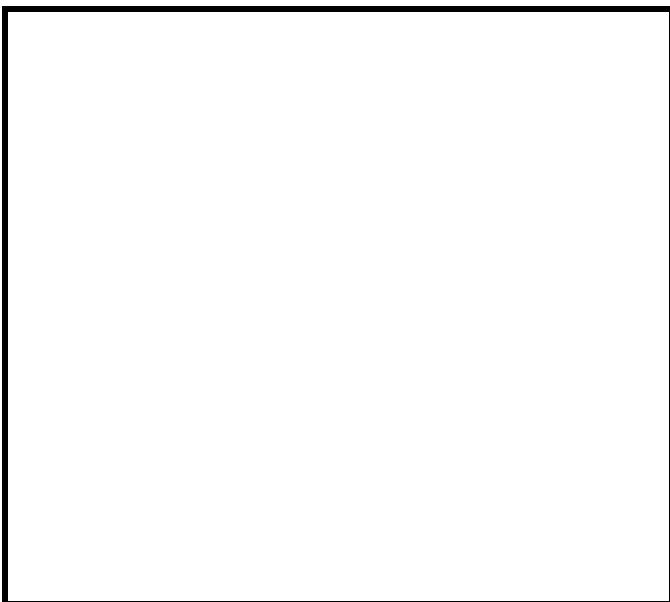
I still remember when Debra emailed the Alliance in early 1999 after finding our web site and noticing that we did not have any AGS families in Nevada. The next I heard, Brian had passed away only one day after returning from California for a check-up—he experienced an intracranial bleed and never recovered.

Debra sent a note with their donation that reads: “We have a special place in our hearts for the children with Alagille Syndrome and also their parents. May God bless you in your endeavors to educate others with your Alliance.” We thank you, Debra and Dan, for your thoughts and prayers!

*–Cindy Hahn, Alliance President*



## Shining Stars



**(far left)**  
Sophia Fung, 1 year old, from Richmond, British Columbia, Canada.

**(left)**  
Justen Angel Giron, 2 years old. This photo provided by his Grandmother, Lorrie Ricci, in Chatsworth, California.

## Cord Blood Donation A Reality at WACCBB

The Summer 2001 issue of *Trail News*, the newsletter of the Oregon Trail Chapter, American Red Cross, included an article about the Western Area Community Cord Blood Bank (WACCBB for short).

The WACCBB, which was started in 1999 as a subdivision of Red Cross Biomedical Services, collects and stores cord blood (blood that remains in the umbilical cord and placenta after the birth of a baby) donated by parents in a region that extends from Portland, Oregon, to San Diego, California. The blood is frozen in a minus-200 degree Celsius freezer until it is needed. Cord blood stem cells can be used in place of bone marrow stem cells in patients with blood diseases. The stem cells migrate to the bone marrow and begin creating healthy blood cells, thus enabling patients to reconstitute

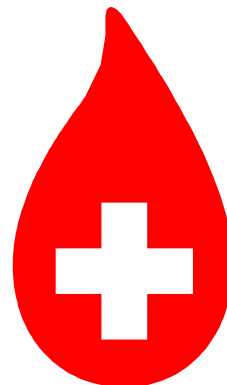
their immune systems.

So far, the WACCBB has shipped only three units of cord blood, all to patients with blood diseases—a 6-year old girl with leukemia in Michigan, a patient with Hodgkin’s disease in Minnesota, and to Massachusetts where it will be used in an experimental procedure to attack a bladder tumor.

On an added note, stem cells also can be used to treat other than blood diseases. For example, researchers at the Oregon Health Sciences University (OHSU) successfully used stem cells from bone marrow, called hematopoietic cells, to repair liver damage in mice that was caused by genetic disease (*The Oregonian*, November 22, 2000, *Science* section). Once purified, then transplanted into the mice, the stem cells went on to form healthy liver cells; according to

Dr. Markus Grompe, OHSU professor of pediatrics and molecular and medical genetics.

Stem cells appear to hold great promise in treating and, hopefully someday curing, numerous genetic disorders. Donation and storage of cord blood is one way of obtaining stem cells that is completely voluntary on the part of the parent(s) and does not carry the moral questions of other sources such as embryos discarded in the in vitro fertilization process.



Anyone who is interested in donating cord blood to the WACCBB, or would like more information about the Red Cross Cord Blood Program in other parts of the U.S., should call Linda Goertz at 503.284.1234 ext. 144, or logon to the Red Cross web site at [www.redcross.org](http://www.redcross.org).



## Trips, Inc. Special Adventures for Special People

Trips, Inc., based in Eugene, Oregon, “organizes trips for people with developmental and other disabilities who need staff assistance for a safe and enjoyable vacation.” Founder and Executive Director Jim Peterson started Trips, Inc., in 1991. Since that time the service has provided almost 300 vacations to over 3,600 travelers from more than 40 states.

The 2001/2002 program includes trips to distant destinations like Scotland and Hawaii, as well as to numerous popular U.S. destinations such as Disneyland, the Grand Canyon, and Washington, D.C.

Call 1.800.686.1013 for a brochure, or logon to the Trips, Inc., web site at [www.tripsinc.com](http://www.tripsinc.com) for more information.

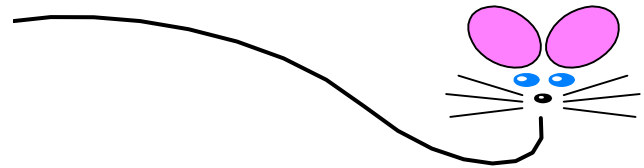


## Camryn Newby Featured on Hugs and Hope Club Web Site

Camryn Newby, who is one year old and has AGS, has her picture and just a bit of her story on the Hugs and Hope Club web site at [www.hugsandhope.com/camrynnupdates.htm](http://www.hugsandhope.com/camrynnupdates.htm). Her mom, Amy Newby, says that she and Camryn's dad, Craig, were told when Camryn was six weeks old that her case of AGS was "terminal". After four days in the NICU, Amy and Craig were told to "take her home and love her" and that's what they have done ever since! According to the Hugs and Hope Club story, Camryn's mom and dad "believe that's what she is living ON—love!"

At one year of age, Camryn weighs just 11 pounds and is very tiny. She loves to be held. Her two sisters, Mikena who is 8 and Karlee who is 5, don't have AGS.

You can email Amy and Craig at [fourplussmiracle@cs.com](mailto:fourplussmiracle@cs.com) (what a terrific address!) or write to Camryn, Mikena, and Karlee through the Hugs and Hope Club at 4550 Horsehead Lake Road, Harshaw, WI 54529.



## J & J : AGS from a Real Life Perspective

Dear J & J,

My daughter Alaina is 11 years old and has AGS. She goes to a week-long resident summer camp in Washington state for kids with chronic illness who are patients at Children's Hospital and Regional Medical Center in Seattle, WA. She's attended this wonderful camp since she was 5 years old and loves it! We were wondering if you know of any other summer camps elsewhere in the U.S. for kids with chronic illness. Alaina would like to travel, but needs the high counselor-to-camper ratio of "special needs" camps.

Thanks,  
Cindy and Alaina Hahn

### Joe says....

It's amazing how a little education and sensitivity can go a long way. This summer I took a Public Speaking class and used one of my oral presentations to help explain some of my own "weird" behavior (scratching/restlessness). On Monday however, I was taking a test in my Algebra class and didn't quite realize how out of control I had gotten until was asked by a student "Is something wrong?" which to me is just a polite way of saying "Stop it! You're bothering me!" It's not really teasing but it's just as embarrassing.

Anyways, as far as camps are concerned, I don't know of any personally but I would encourage you to check out [www.kidscamps.com](http://www.kidscamps.com) which would be a good starting resource. As a director for the AGSA I would love to see a summer camp established for AGS kids. It is something I plan on working on and hopefully make a reality in the not-so-distant future.

Look forward to hearing from you again....

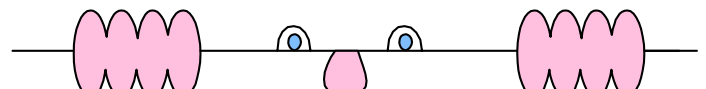
Dear J & J (and AGS Parents on the Alliance BB),

My daughter who is 2 next month has AGS and as with many, her main problem is itching! Her sleeping patterns are usually about two hourly (during which time she is constantly scratching) - then she wakes up frustrated and can't go back to sleep. How do parents of kids with AGS or AGS sufferers cope with all the sleepless nights?

Sincerely,  
Lisa

### Justine says....

I honestly don't have a clue how I manage to sleep at night. What helps though (although it won't help your daughter yet) is that I relax (as much as I can that is) and keep telling myself not to itch. I wake up a lot during the nights and that's when I have the most trouble getting back to sleep. I just do the same thing as before but it takes longer to get to sleep.



Email J&J at: [JandJonline@earthlink.net](mailto:JandJonline@earthlink.net)

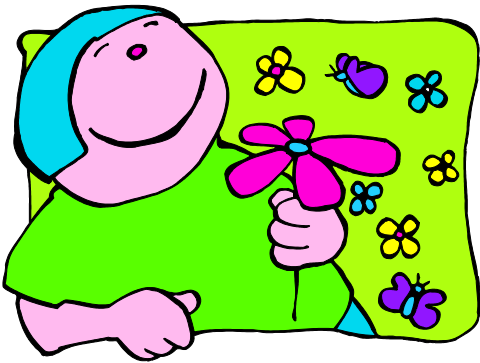
or

Respond to their questions on the Alliance Bulletin Board at [www.alagille.org](http://www.alagille.org)!



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Email: alagille@earthlink.net

**WE'RE ON THE WEB**  
**WWW.ALAGILLE.ORG**



## Symposium 2002 Update

The Second International Symposium on Alagille Syndrome—or S-2002 as the Alliance has taken to calling it—will definitely be at Eastern College in St. David's, PA, August 8-11, 2002. Mark your calendar and plan to join us!

**Great news about Childcare and the Kids' Program...** The Alliance will be hiring a destination management company called Even-tions to coordinate the childcare and Kids' Program for S-2002. This company is fantastic!

They will provide toys, supplies, and childcare providers for the little ones, as well as counselors and equipment for entertaining the rest of our kids. All childcare and kid's programs will be provided on site at Eastern College, so children will be well cared for and close by while parents are in sessions. Events also will assist at the Friday Luau and the Saturday Ballpark BBQ with some special entertainment for the kids.

**A special thanks to AGS families who**

responded to our invitation to become "Symposium 2002 Friends" by making a donation of \$250 or more toward the S-2002 fund at this time. As of this writing, the Alliance has received \$3,900 in donations to help cover the cost of the down payment to Eastern College to hold the S-2002 dates. We greatly appreciate your assistance and hope to see you at S-2002!

Look for more information about S-2002 in the next *LiverLink*.

## Symposium 2002 Preliminary Program Highlights

### Thursday, August 8

**AGS 101**—The Basics, a workshop for people new to AGS (two sessions)

**Welcome Reception & Conference Kick-off** "Celebrating Community", making Community Quilt Squares, Pizza Buffet & Ice Cream Sundaes

### Friday, August 9

**Keynote Address**—Parenting Children with Medical Challenges

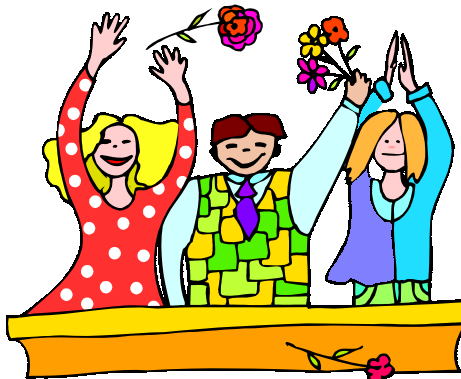
**Family Panel**—Coping in a World Gone Berserk: Adjusting to the AGS Diagnosis

**Breakout Sessions Galore**—School Issues, Special Needs Estate Planning, Adult Issues, Alternative Therapies, and more

**Keynote Address**—The AGS Research Agenda and the Significance of Notch/Chromosome 20

**"It's a Luau"** Hawaiian Beach Party and Shish Kebob Buffet

**Evening Forums** for Teens, Young Adults, Dads, and Moms



**Fun**

### Saturday, August 10

**Keynote Address**—The Implications of Stem Cell and Genetic Research Advances for AGS Diagnosis and Treatment

**More Breakout Sessions**—Nutrition, Lipids & Xanthomas, Cardiology, Growth Hormones, Dental Issues, Vision, Itching, and much more

**Group Pictures**

**"Let's All Go To The Ball Game"**

Ballpark BBQ Party

**Evening Forums** (Repeat)

### Sunday, August 11

**Keynote Address**—Courageous Coping: Living with a Chronic Illness

**"Tell Us Your Story"** for Families

**Conference Closing & Brunch Buffet**, making a Community Chain

## AGS for Kids: A Story for Alaina's Classmates... by her Mom, Cindy Hahn

1

"Alagille Syndrome—it's a liver disorder, which means Alaina's liver doesn't work right. She was born with it, but we don't know why it picked her and not someone else."

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"Does anyone know what jobs your liver has to do in your body? Well here is a list of just a few of its 5,000 jobs!"

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"Alagille Syndrome, or AGS for short, isn't just a liver disorder. It also affects a lot of other parts of the body. This picture shows those parts."

4

"One of the worst effects of AGS is itching. Sometimes Alaina itches so much, she feels like a cat gone crazy! She even scratches in her sleep!"

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"Sometimes Alaina feels like an alien, because she is different from other kids and she rubs her nose a lot and gets hives when she has a cold."

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"Alaina gets frustrated and angry about being different, especially when other kids make fun of her and when she feels left out."

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"Sometimes Alaina feels like she has a target painted on her shirt that tells kids, 'Go ahead and tease me.' She feels like kids don't listen when she tells them she can't help being different."

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"All Alaina wants to be is a normal kid, like everyone else. She wants other kids to like her and to accept her as she is, even if she has AGS."

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"Does anyone have questions? Alaina and I would be glad to answer them as best we can!"