

LiverLink

The Newsletter of the Alagille Syndrome Alliance

The 7 Year Itch by Dr David Piccoli and Kathleen Loomes

Special points of interest:

- July 14th-AGS/Gehman Picnic in Philadelphia, PA
- Got an itch?? Check out the article from Dr Piccoli on itching.
- [Who are J & J.](#) Find out in this issue
- [Which way did we go??](#) [Check out the President's Page](#) to find out where the Alliance is headed.
- [Symposium 2002](#)-It's not just a date on a calendar.

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Although itching (pruritus) is not one of the dangerous manifestations of Alagille Syndrome (AGS), it certainly is one of the most common and vexing problems. This itching typically worsens in the first years of life, and then for some patients progressively improves. In others, itching is a severe annoying problem that can last for many years, despite a liver that otherwise is functioning well. Sometimes the itching is mild and seasonal, causing no real interference with normal activities, but in some children it is so severe that it interrupts sleep and causes constant bleeding and scabbing. Rarely, itching has been the indication for major surgery or even liver transplantation. Some school function and attention problems are undoubtedly due to chronically interrupted sleep, and inattention and distraction due to the constant itchiness dur-

ing classes. Itching is not a problem associated with all forms of liver disease, but it is a particularly important part of AGS.

The reasons for this itching and the pathways involved in producing itching are still not well understood. The itching in liver diseases appears to be related in some significant way to the bile salt levels in blood and skin, but this is not the entire answer. The sensation of itching can be mimicked by medications like morphine or Demerol, which have a direct effect on brain pathways producing itching. This effect can be blocked at the central (brain) level by using opioid receptor antagonists (such as naloxone and naltrexone), which block this itching pathway. For people (without AGS) who have allergies, the itching eyes, nose, throat and skin can be treated very successfully with antihistamine medica-

tions. Newer generations of these antihistamine medications have been developed for allergy sufferers who can achieve great symptom control. These antihistamine medications are used in infants and children with liver itching, but the results are much more modest. Damage to the skin itself, whether caused by an insect bite or by a scab, can result in local itching. Sometimes a significant improvement in symptoms can be achieved by treating the skin directly. This type of therapy is used in AGS, again with somewhat variable results. Finally, if the bile (or bile components such as bile salts) which are poorly excreted in AGS are the cause of itching then strategies which would decrease these components in blood and skin might be successful.

[Continued Page 3](#)

Family Events

Alagille Picnic - July 14, 2001 -

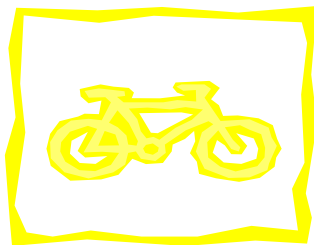
Get out your supersoakers and head to the Gehman's for the annual Alagille Picnic on July 14! It's sure to be full of soaking wet fun for the whole

family.

Be prepared to learn something new about AGS, make new friends, and get reacquainted with families from past years. Staff from the Children's Hospital of Philadelphia (CHOP) really has a

blast at this event and so will you!!

For information, contact Lowell and Teena Gehman at 1840 Oaklynn Drive, Green Lane, PA 18054; phone: 215.538.3672; email: lgehman@excite.com.



President's Page

Good Day, Sunshine! I can't help feeling pumped about the Alliance these days. I'm still feeling the elation of our first ever Board of Directors Retreat on May 4-5 here in the Rose City (Portland, OR). Our dedicated team of volunteers – Patti Everett, Joe Anderson, Julie Kelin, Beth Caldwell, Erik Luxhoj, and Chris Hahn – gathered for the event and came away energized and excited about the Alliance and its future.

During the two-day Retreat, we set short- and long-term goals, launched into planning for Symposium 2002, and identified ways to update and improve our Web site – with lots of good humor to boot! We also were able to get to know one another as a Board and develop a sense of shared mission – or as Joe Anderson would say “we bonded” – that will propel us in the next few years as a unified team, helping one another to accomplish our vision and goals to the benefit of the Alliance and AGS community. A summary of our notes from the Retreat is included in this LiverLink. Take a look and see how high we've set our sights! If you see a role for yourself in our planned activities, please let us know. We

welcome your comments and suggestions, as well as your participation in making our vision and goals realities.

The site and dates are set for Symposium 2002! We've chosen Spring Mountain Camp, Retreat & Conference Center in Schwenksville, PA, for June 13-16, 2002. Located on 300 acres in the foothills of the Pocono Mountains, Spring Mountain is only 45 minutes from Philadelphia, PA, and 2 hours from New York City, NY. It's a beautiful place, with amenities galore that can accommodate up to 400 people. On-site accommodations include a lodge that sleeps 50 people, seven “villages” with 7-10 cabins each (all with private bath facilities), and a campground. Horse stables, a bike trail, hiking trails, a pool and private lake, athletic fields, recreation halls, and a giant water slide are only a few of the numerous facilities for family fun that Spring Mountain offers. We're planning to run the conference from Thursday dinner to Sunday brunch, packed with family activities, sessions on AGS and other topics of interest, and a kids programs/camp, with a summit for scientists involved in AGS research

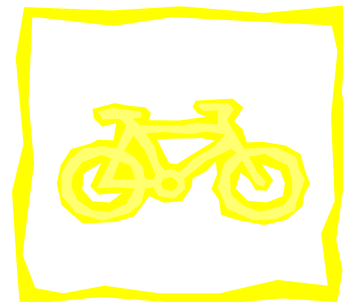
running concurrently. Mark your calendar and plan to attend! Details will be published in future issues of LiverLink.

I hope you find the Alliance Financial Summary for 2000, which is included in this issue (see Page 4), informative. As you can see, our main source of income is contributions from AGS families, and our biggest expense is printing and mailing of LiverLink and information packets for new families. If you have any questions about the summary, please feel free to contact me. We welcome your comments.

Enjoy the summer and stay cool!

Until next time, keep smiling and be well,

Cindy L. Hahn, President



“...we set short- and long-term goals, launched into planning for Symposium 2002, and identified ways to update and improve our Web site –”

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. It's 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©2001Alagille Syndrome Alliance. All rights reserved.

The 7 Year Itch (continued)

Agents such as cholestyramine are designed to bind bile salts in the gut so that they cannot be reabsorbed. Another approach would be to drain the bile externally (for instance, to the skin) so that it could not be reabsorbed in the bowel. This therapy is extraordinarily successful for some diseases other than AGS. There is more limited success in AGS patients, due in large part to the fact that many patients during their most intense itching years, can actually secrete very little bile into the biliary system, therefore leaving little to be drained externally. Other approaches, including those to breakdown bile salts in the skin, such as ultraviolet light treatment, have also been used to treat the itching from liver disease.

Despite all of these potential therapies, itching remains the most important symptom associated with AGS, and more work needs to be done to improve pruritus in our patients.

Therapy for Pruritus

Non-absorbable ion exchange resins — cholestyramine (Questran) and colestipol.

These medications are resins which bind bile acids, cholesterol, many drugs, vitamins and other potentially toxic agents. These compounds bound to the resin are then excreted in the feces. These resins will decrease the reabsorption of bile acids, which is a normal beneficial process in people without itching and without liver disease. If they bind a significant amount of the bile acids, the body will be stimulated to turn more cholesterol into new bile acids. In patients who excrete a significant amount of bile, this can be an extraordinarily beneficial effect. These medications were developed to decrease cholesterol levels using this same pathway and mechanism. Unfortunately, these resins can bind important medications, they do not taste particularly good, and they can be difficult to successfully administer. It is important that other

medications and vitamins not be given within about two hours of these resins. Finally, many patients just do not seem to get significant beneficial effect from these medications.

Rifampin (rifampicin)

Rifampin is an antibiotic that was developed for tuberculosis therapy. It was shown to be very effective for some patients with adult cholestatic liver diseases and the experience in pediatric patients generally has been good. A significant number (but certainly not all) of patients will have a dramatic reduction in their itching when they are treated with Rifampin. There are now several studies that have shown a beneficial effect in children. The precise mechanism for the action of rifampicin is not known, but it does appear to directly affect the liver cells' uptake and metabolism of compounds including the bile salts. It does not appear that the antibiotic activity of Rifampin is involved in its effect on itching.

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Please let the Alliance know if you move or your email address changes! Your help in keeping our records current is appreciated.

Internet Connections: Band-Aids & Blackboards- Leaping Good Fun

Don't waste another minute – logon to Band-Aids & Blackboards at <http://www.faculty.fairfield.edu/fleitas/contents.html> for hours of feeling connected to chronically ill kids, teens, and adults! In the words of its creator, Joan Fleitas, RN, Associate Professor of Nursing at Fairfield University in Fairfield, CT, this site is “about

growing up with medical problems...any ole type.” Band-Aids & Blackboards goal is “to help people understand what it's like, from the perspective of the children and teens who are doing just that.”

Full of eye-catching graphics and super user-friendly, this site draws you in and keeps kids connected with

topics like: Why do kids get sick?, All about teasing, A daydream for sick kids, and Pranks you can play in the hospital. Teens have similar intriguing topics from which to choose: Stories from the front; The good, the bad and the ugly; Experts share their tips; and Poetry speaks truth. Parents and teachers can glean valuable information

from the Adult Frog Pond on: Sibling relationships, Adults remember when (they grew up with a chronic illness), Inclusion, Resources for teachers, and The lowdown on teasing.

Check it out! You'll no doubt keep coming back for more!

Financial Update**ALAGILLE SYNDROME ALLIANCE
FINANCIAL SUMMARY
JANUARY 1 – DECEMBER 31, 2000****Income**

| | |
|--------------------------------|-----------------|
| Contributions | 5,698.00 |
| Investment Income (Interest) | 114.75 |
| Other Income | 432.00 |
| Total Income Categories | 6,244.75 |

Expense

| | |
|---------------------------------|-----------------|
| Affiliations | 80.00 |
| Bank Charges | 55.14 |
| Charitable Donations | 20.00 |
| Copies | 109.76 |
| Corporate Fees | 10.00 |
| Fax | 4.00 |
| Mailing (including printing) | 2,246.85 |
| Miscellaneous | 50.00 |
| Postage | 186.60 |
| Promotional | 535.25 |
| Supplies | 524.24 |
| Transportation | 24.00 |
| Utilities | 223.92 |
| Total Expense Categories | 4,069.76 |

Income vs. Expense **2,174.99**

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 soar above the clouds!

The 7 Year Itch(continued)

Rifampicin does appear to be safe for long-term use, but it should be carefully monitored. In general, this is a good agent that can have dramatic results.

Antihistamines

Antihistamines are medications that block the action of histamine in the body. Histamine in different organs can play dramatically different roles. In one pathway, histamine is involved in stomach acid secretion, and the “H2-histamine blockers” are the medications that have been successful in blocking stomach acid, and treating reflux and ulcer disease. Histamine is also involved in itching. Many infants with AGS have been treated with Benadryl (diphenhydramine) or Atarax (hydroxyzine), or related compounds. These medications may cause sedation, and in some children there may be a paradoxical effect. Generally, the sedative properties of these medications become inapparent after a few days of therapy. Although antihistamines work well for purely allergic itching, the results in AGS have been more variable. Some parents find that a larger dose at bedtime, which has the sedating effect, is more beneficial for their children than round-the-clock medication. As these medications are used over a long period of time, children can tolerate higher and higher doses without apparent side effects.

Phenobarbital

Phenobarbital is another complicated medication that is used as an anti-seizure medication and also as a sedative. It appears to work for some patients with itching by increasing the bile flow, and it causes complicated metabolic changes in the liver. Phenobarbital was a very common therapy for jaundice and liver disease in the past, but its use more recently has been limited, as studies comparing its effect to other agents have not shown it to be as beneficial. Phenobarbital can also have some important metabolic effects on vitamin levels and careful monitoring is necessary.

Ursodeoxycholic Acid (Actigall, Urso, ursodiol)

Ursodeoxycholic acid is actually a favorable bile acid that improves bile flow. It has been shown to decrease itching in many different liver diseases, including AGS. It probably functions both by improving bile flow and by changing the composition of bile and the body’s bile salts. However, if bile flow is dramatically limited, particularly in infancy and the early toddler years, ursodeoxycholic acid therapy may actually lead to a significant worsening of itching, and there are times when stopping this therapy for a few weeks is a necessary biological experiment.

[Continued on page 8](#)

Wish Upon a Star/ Starlight Foundation

Most people have heard of the Make-A-Wish Foundation and its wish-granting to terminally ill children. Well there’s another organization that grants wishes to sick children, but they can be chronically ill or medically fragile rather than termi-

nally ill. It’s called the Starlight Children’s Foundation and you can find information about it at <http://www.starlight.org>.

Besides wish-granting, the Starlight Children’s Foundation sponsors Starlight Sites,

Starlight Fun Centers, PC Pals and Pal Net Planet, Hospital Happenings, and Kids Activity Network excursions. These programs are designed to provide opportunities for children “to escape the drudgery of illness” and forget about the

pain, if just for a while. The Foundation is based in Los Angeles, CA, and has local and regional chapters serving various states in the U.S.A. and countries abroad.

Logon and start reaching for those stars!

News Clips

Brain Chemistry and Vitamin A Linked

According to researchers at the Salk Institute and the National Institutes of Health (Bethesda, MD), Vitamin A deficiency impairs brain chemicals important in learning, both in mice and adults. Studies on mice, which were deficient in Vitamin A for at least 15 weeks, found that chemicals related to brain-cell adaptability were impaired as a result of the deficiency.

Both LTP (long-term potentiation) and LTD (Long-term depression) mechanisms, believed to be responsible for learning and memory in the mouse brain, were impaired. The damage, however, can be reversed in as little as two days by restoring Vitamin A to the diet.

Sharoni Jacobs, a Salk graduate and researcher, said that "There's an obvious significance to this for humans suffering from Vitamin A deficiency." Another re-

searcher, Ronald M. Evans, noted that the study indicates "the detrimental effects of Vitamin A deprivation are remarkably reversible," offering hope to those suffering from Vitamin A deficiency.

Source: Adapted from "Study links vitamin A, brain chemistry" by Jeff Ristine, San Diego Union-Tribune, fall 2000.

Birthdays!!!!!!

January 2001

Dalton Baker 6
Caley A. Corbett 10
Tessa J. Derusha 9
Jeffrey Dubrawski 14
Benjamin T. Earnshaw 3
Luke T. Fasano 9
Lauren Gajdosik 10
Kayla A. Garland 11
Heather L. Hansen 18
Jesse L. Isabelle 9
Brianna James-Beckham 7
Mitchell Jelley 13
Megan Lavallee 5
Russell W. McCoy 26
David M. Rader 2
Hannah L. Zinno 5

February 2001

Samantha L. Armstrong 19
Kasie F. Brown 18
Benjamin Doehling 10
Zachary Flores 8
Carine Hendriks 5
Connor Quillen 2
Trevon "Tre" M. Rountree 2
Kendall N. Shepard 2
Kristopher R. Stanley 11
Tara Tchalabi 23

Tara Tchalabi 23
Kate Thomas 7
Jessica Wood 10
Sheena Wood 15

March 2001

Ashley C. Boyce 2
Jose L. Campos, Jr. 12
Dylan K. Dignam 5
Ross Garrison 10
Ali Gorham 5
Cole Harden 18
Rachel Herschmann 3
Susan Heuser 43
Anna M. Laurent 6
Amanda Marx 8
Helen Ouellette 16
Ernest Ouellette 13
Christopher J. Smith 12
Anthony Strukel 3
Leo G. Westwind 6
Thomas Whyte 4
Sheryl M. Zook 20

April 2001

Alexis Baldwin 6
Ryan Dunk 1
Sandy King 32
Jed McConville 6

May 2001

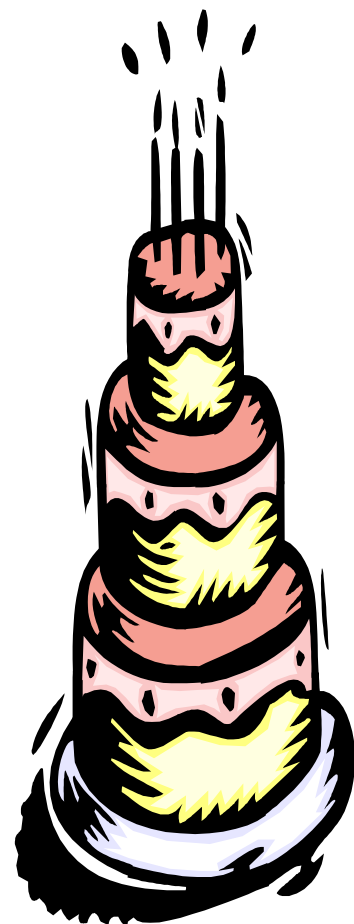
Christopher J. Ciesiel 6
Colten R. Gibbs 8
Joshua J. Heinke 14
Danielle E. Nathan 6
Matthew E. Neininger 4
Roxanna Pakravan 5
Jordan K. Sihle 8
Nicholas Tilger 5

June 2001

Iain A. Barr 7
Michael Blais 15
Gregory Bryant-Bruce 8
Claudia J. Chung 6
Krista Doornenbal 8
Ryan Duncan 14
Neema Khazaie 5
Angeline R. Macias 1
Jean K. Marchel 43
Justine Sawaya 14

In Memory

Sylvian R. Briegoos 7
Alyssa M. Parker 6
Shelby N. Butler 5
Hayden Gibson 10
Carly Rosendal 6



And Now—A Word from Your Board

Excited! Committed! Energized! These are the words that describe the Board of Directors after their Planning Retreat in early May. The Board met with Diane Tutch, a planning consultant, to create a plan for the short- and long-term future. The expected outcomes were:

- To create a vision for the future, with a picture of how the organization can grow
- To assign responsibilities to each Board member, working toward that Vision
- And to develop objectives for one, five, and ten years in the future to guide the Board.

This helped the Board form a vision statement:

In ten years the AGSAlliance will be the primary contact point for people with Alagille Syndrome (AGS), their families, and friends. Members will form a close-knit community, gathering together to provide mutual support, information, and tangible assistance. This community will be facilitated through the Internet and through active regional

groups. The AGSAlliance will bring the entire community together every three years, and the regions will be encouraged to gather annually.

The AGSAlliance will be a source of tangible support for people with AGS, their families and friends. Such support may include hosting an annual camp for AGS children and young adults, offering college scholarships to people with AGS, and providing guidance in obtaining insurance coverage and health services. In the long-term, providing emergency assistance to families in crisis may be added as a support service. The AGSAlliance will be the source of current, accurate information on AGS and its treatments. The organization will be a financial supporter of research and will be actively involved in encouraging its members to participate in research studies. The AGSAlliance will be a source of referral information, maintaining information about physicians and medical programs

offering effective treatments for AGS. The AGSAlliance will be a source of information for health care providers seeking to initiate or enhance their services to people with AGS.

The AGSAlliance will be a well-run organization with paid staff providing effective services. The organization will be supported through a well-established fundraising program using a variety of techniques to generate funding to support the priorities of the Board of Directors.

Using the goals as a starting point, the Board began planning ways to make the vision statement a reality.

One of the first goals we tackled was that of our support network. It was decided to update the Web site at www.alagille.org using a new format and host, creating a message board for support, and a new feature, called “Ask J&J” for kids, teens and young adults with AGS. The Board is very excited about the role the Web site will play in the future of the Alliance.

We next looked to identifying regions, with regional coordinators. In the future, once established, each regional group would be able to distribute information, welcome new members, plan regional events, and submit information for the Web site. It is our goal that the Symposium will eventually be held every three years, on a rotating basis in each region, and that each region will have had one regional event by the year 2006.

The Board is also creating some brochures. The first one, to be completed by the end of summer, will be for teachers, therapists, and other lay people who need concise, correct information about AGS.

The rest of the retreat was dedicated to planning for Symposium 2002. See the President’s Page for details!!!!

Helpful Hints

Gulp! It’s a Pill!

If your child has a hard time taking pills, or just downright refuses to consider the idea, here is a strategy that might work.

First, show your child how to take a pill by swallowing one yourself. If you don’t take pills, use a fake one – an M&M or a jellybean. Put the “pill” in the back of your mouth, keeping your tongue flat, then take a mouthful of

water, tilt your head back slightly, and swallow. Voila! The pill is on its way to work. Now have your child give it a try. Start small with something like a cake decorating sprinkle (very small, oblong, colorful). Gradually progress to bigger and bigger “pills” allowing your child time and repetitions at each step until he or she is comfortable and swallows the pill easily. Suggested “pill” sizes, from small to large include:

1. Cake decorating sprinkle
 2. Silver cake decorating ball (small, shiny, silver, seem to roll all over the counter)
 3. Round, multicolor candy (Spree or M&Ms come to mind here)
 4. Red licorice whip cut to 0.5-inch length
 5. Capsule-shaped candy, multicolored (such as Tic Tacs or Dynamints)
 6. Normal-sized capsule (child’s actual pill)
- Happy pill swallowing!

Source: Adapted from “A brief, effective method for teaching children to swallow pills” by R.L. Blount, L.M. Dahlquist, R.A. Baer, and D. Wouri, 1984, Behaviour Therapy, 15. p. 383. Provided to the Alliance by Erika Byers, RN, FNP, a labor and delivery nurse at the Oregon Health Sciences University in Portland, OR.

Letter Box

Tina Valente (Tucson, AZ) reports that **Joey's** cholesterol decreased from 600 to 238 in the 2 months after he started taking Lipitor. His itching also improved significantly. Susan Krug, MS, RD, at the Children's Hospital Medical Center in Cincinnati, OH, thought that the reduced itching could be due to reduced irritation in his skin from "pre-xanthoma" activity as the cholesterol has decreased; this is a theory she suggested in a telephone conversation with Tina. Tina also wanted to know if anyone has heard about the use of Zophran for itching. An article in *The European Journal of Pediatrics* (Vol. 155, p. 990, 1996) discussed a two-year-old patient with AGS who was treated with the drug and experienced reduced itching.

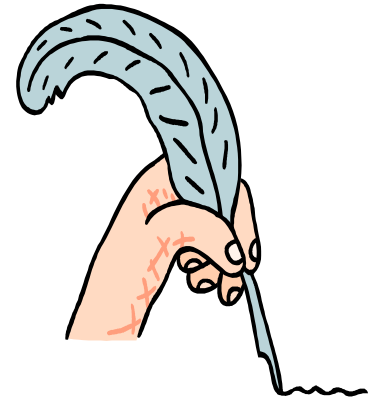
Terri Dragony (Tucson, AZ) writes that she had to pull **Jacob** out of school temporarily due to an "overall lack of supervision" by the school district, which was putting him at risk of a head injury. She has since discussed the situation, at length, with the district. As a result, Joey now has a full-time aide and can come out of Special Education to be fully mainstreamed with other children his age. Terri noted that integrating AGS kids into the public school system can be difficult. Most of the children are intellectually fine, but medically fragile, and school officials and staff are not sure how to accommodate them.

Evelyn Cannon (Norcross, GA) informed us that the **Walker Cannon Memorial Chapel** is completed and "is an awesome and peaceful place." She writes that the Chapel is beautiful and "will honor the wonderful life of our special child." The Chapel dedication was held on Sunday, February 4, 2001.

Patti Everett (Alpharetta, GA) writes that **Alex** is in pre-Kindergarten and is happiest when he is coloring, singing country songs, and just clowning around. He had a feeding tube placed in September 2000 and now seems to be gaining weight and energy with nightly feedings. Patti says that Alex "just seems to take life and enjoy it as it comes" and can be counted on to "help us remember what is truly important" in life.

Terrie Dignam (Federal Way, WA) reports that **Dylan** has grown quite a bit and "is acting very four!" He goes to school four afternoons a week and has speech, occupational, and physical therapy both in and out of school. In 2000, Dylan underwent bowel obstruction surgery and brain surgery to improve blood flow and help prevent future stroke activity. The Dignams also traveled to Chico, CA, for six weeks of Hyperbaric Oxygen Therapy for Dylan. He's shown improved gross and fine motor function, better clarity of speech, and better overall balance as a result. They hope to continue these treatments this coming summer.

Willem Verburg (The Netherlands) writes that daughters **Mariska**, **Martine** and **Tilly** have experienced decreased itching after being put on Nalorex (125mg twice a day). It seems to be most effective when combined with Rifadin (Rifampicine; 300mg twice a day). Martine and Tilly (twins) are now able to sleep at night and are very happy to be "living a normal life." Mariska still experiences more itching than her sisters because she also has biliary atresia. Willem encourages other parents to ask their children's doctors about Nalorex and hopes it will help other AGS kids with severe itching.



"Joey now has a full-time aide and can come out of Special Education to be fully mainstreamed with other children his age."

Symposium 2002!!!!!!!!!!!!

YES! Planning for Symposium 2002 is well under way! Next year's Symposium will be held the weekend of **June 13-16**, at Spring Mountain Camp in Pennsylvania. Spring Mountain has a Web site at www.SpringMountain.com if you would like to visit there.

We have already lined up several GREAT speakers, and plan on having an even better time than at the last Symposium! Spring Mountain is a wonderful facility that would make combining a family vacation with Symposium 2002 a lot of fun! More info will follow!!

Our Internet Connection!!!!

Exciting news! Under the capable management of Erik Luxhoj, our Internet site, www.alagille.org, is undergoing some major changes. When completed, the site will contain a research library, pamphlets for teachers or

other professionals, Ask J&J, a support page for AGS kids, teens, and adults, and most exciting of all, a **Bulletin Board** for everyone to post questions, answers, and general updates to. We can't wait for Erik to finish these updates!!!

The 7 Year Itch(continued)

Opioid Antagonists

Because there is now good evidence that itching is “felt” centrally in the brain, medications which block the receptors involved in this itching can be successfully used to treat pruritus. Naloxone is a potent opioid antagonist that can be given as an infusion, but it has certain chemical properties which do not make it a good oral medication. Naltrexone is an opioid antagonist which can be given orally, and some patients have substantial success with this medication, although a number of patients may have side effects which, although not dangerous, show an intolerance to the medication. Longer term and pediatric trials are certainly necessary before we can fully understand which patients are appropriate for this class of medications.

Other Therapies

Tegretol (carbamazepine) is an anti-seizure medication that has been shown to reduce itching in some patients with liver disease and it also has been shown to decrease pain in certain neurologic conditions. Unfortunately, one of its side effects is liver toxicity, which although rare, may complicate the liver disease in AGS. Some patients have used phototherapy with ultraviolet light of either the UVA or UVD type. The results with this therapy have been quite variable, and as yet there are no good studies in pediatric patients or infants. Ondansetron, which is an anti-vomiting agent, and propofol, which is an anesthetic agent, have also been explored as agents for itching.

Local Therapy

It is important to emphasize that the skin itself is damaged by itching, and that damaged skin can be a stimulus for more itching. This is commonly recognized (in the absence of liver disease) when a scab is in the healing phase and it stimulates an itchiness that, unfortunately, commonly causes it to be “picked open.” Furthermore, the scarring that is seen in patients with AGS is due in part to the constant breaking of the skin by the fingernails or other objects which are used to itch the skin. Some patients have dramatic results with using a cream or ointment on the skin, particularly at night, to improve the integrity of the skin. Fingernails should be clean and well-trimmed. Some patients cause substantial destruction of their skin during sleep, and cotton gloves worn at night may help substantially. It is also important that bathing times be limited, and that strong soaps not be used, as these can have a very significant drying effect on the skin, further exacerbating the itch cycle.

Surgical Therapy

It is possible surgically to divert the bile externally in order to interrupt the cycle of the reabsorption of bile salts and other bile components back into the body. This therapy has been extraordinarily successful for patients with some other forms of inherited liver disease. The results in AGS have not been as dramatic, although some patients

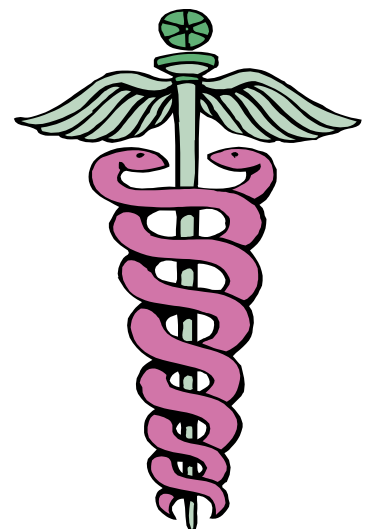
have achieved some significant success. More extensive surgical therapy, meaning liver transplantation, is quite effective as a last resort for severe intractable pruritus, and at times it may be necessary to consider transplantation despite the fact that the liver otherwise is functioning well. This is obviously a complicated decision that should be made with your child’s liver doctor, as well as with a transplantation center.

Long-Term Course

In most patients with AGS, the itching progressively worsens during the time course when the bilirubin, cholesterol, bile salts, xanthomas, and jaundice all worsen, and then subsequently improves and in most patients eventually resolves. It is important to consider, therefore, that the therapy for itching in many patients is a bridge to get to the time when the liver’s excretion function improves to the point where itching goes away. Therapies which are administered during the phase when a patient is already improving may seem to give a dramatic result. It is important, however, that anti-itching therapies be withdrawn occasionally so that parents and doctors can assess whether they were, in fact, actually improving the situation or not. Itching remains the biggest single clinical symptom facing the majority of patients with AGS. Despite ongoing research, a perfect therapy has not yet been devised, and patients respond differently to combinations of

the medications described above. The response to these medications cannot easily be predicted, and commonly it is necessary to adjust different medications at different levels and in different combinations until adequate control of the itching is achieved.

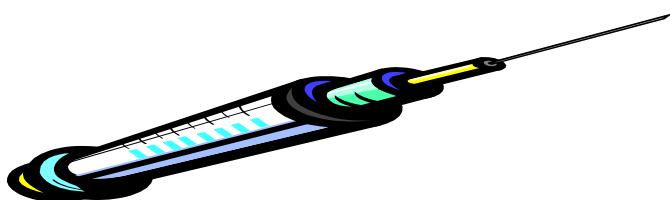
Dr. Kathleen Loomes is an Assistant Professor of Pediatrics at The University of Pennsylvania School of Medicine and Dr. David Piccoli is Chief of the Division of Gastroenterology and Nutrition and Professor of Pediatrics at the Children’s Hospital of Philadelphia (CHOP). The Alliance would like to thank them both for this very helpful information on a very itchy subject.



Sleepy Juice? A Clown Mask?

Just in case you're wondering, sleepy juice is the "kid-friendly" term for anesthesia, and a clown mask is, you guessed it, an anesthesia mask. Two examples of "kid-friendly" medical terms developed by experts at Miami Children's Hospital (published in the fall 2000 issue of *Parenting* magazine) to ease a child's fear when visiting the doctor or a hospital. The idea is that children who understand a procedure or how equipment will be used won't be as afraid as those who hear only medical lingo that is way over their heads.

Here is the rest of the list of "kid-friendly" terms:



| Say..... | Instead of.... |
|----------------------|---------------------|
| Pinch | Shot |
| Arm hug | Blood-pressure cuff |
| Picture | X-ray |
| Camera | X-ray machine |
| String | Band-Aids Stitches |
| Big rubber band | Tourniquet |
| Super Band-Aid | Cast or Dressing |
| Bed on wheels | Stretcher |
| Small medicine straw | IV |
| Small opening | Incision |
| Wake-up room | Recovery room |

Wouldn't it be great if all medical staff used these simple words with kids?

Contribution Cudos (January 1-May 15, 2001)

In Honor of Ben Doehling's Birthday

Ted & Marian Doehling

In Honor of Mary-Genevieve Moisan

Kate & Michael Corrigan

Bob & Ann Osterhaus

Matt & Marilyn Osterhaus

Ray Townsend & Jane Osterhaus

Luke & Jill Osterhaus

Amy & John Moisan

Betsy Osterhaus Hand

Mark Osterhaus

John & Mary Pat Osterhaus

Molly & Bob Whitmore

In Memory of Christopher Minard

Scott & Laurie Minard

In Honor of Patricia Nicole Espin's 3rd Birthday

Barbara Bisgrove

In Honor of Roxanna Pakravan

Firouzeh Fouladi

In Honor of Rachel Herschmann

Eric & Debra Herschmann

In Honor of Megan Lavallee's 5th Birthday

Tom & Laura Lavallee

In Honor of Christian Druckhammer

Kathey & Dennis Druckhammer

In Honor of Trevon M. Rountree

Alma J. Davis

In Honor of Hannah Zinno

Jim & Sandy Hardin

In Memory of Alyssa Parker's 6th Birthday

Stacy & Steve Parker

Barb & Abe Weitzberg

In Honor of Brandon Nicklay

Chuck & Judy Nicklay

In Memory of Walker Cannon

Penny & Tony Pickett

In Honor of Zachary Damir's 4th Birthday

Joel & Dianna Damir

General Donations

Kelli Abadie

Lee & Sam Bandh

Walther & Caecilia Hahn

Margo Pattison

Raymond & Victoria Schrader

Updates.....

Eric Murbach, an AGS young adult, just graduated from Purdue University with a Bachelor of Arts in Communication. Congratulations, Eric!!

Alisha Rovner, a wonderful research coordinator and contact person at the Children's Hospital of Philadelphia (CHOP),

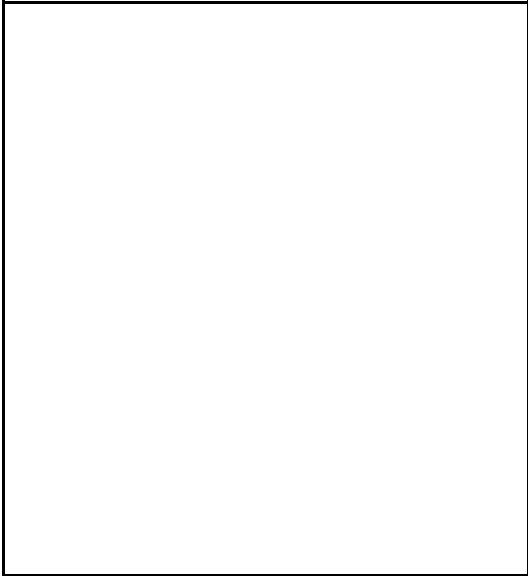
will be leaving for graduate school in Boston. Alisha has been a wonderful support, and we will miss her. She assures us that she will be back for the AGS family days at CHOP, and other AGS Alliance events. Good luck in graduate school, Alisha!!



The Newsletter of the Alagille Syndrome Alliance

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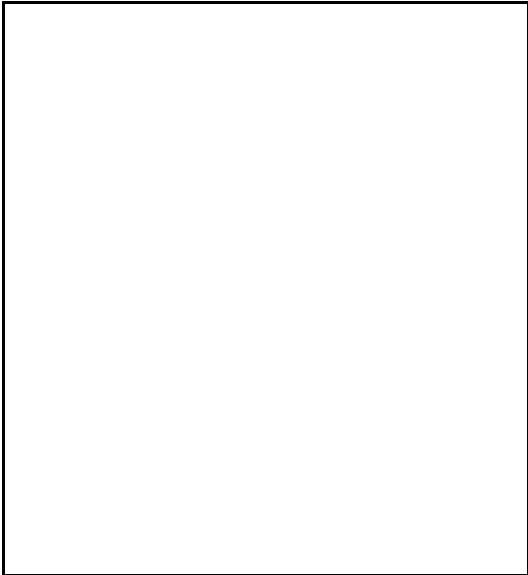
Meet.... Joe Anderson

Meet J.....& J

Hi, my name is **Joe** Anderson and I'm 23 years old and I have AGS. I'm a college student majoring in Finance. I enjoy watching and playing sports and chatting online with my friends. If you have any questions about what it's like to grow up with AGS, let me know and I will try to answer them based on my own experiences and wisdom (or lack thereof)
:^)....

My name is **Justine** Lynn Sawaya. I'm 13 years old & I've had AGS since I was born. I live in San Diego, CA & I'm in the 8th grade - about to graduate from middle school. I like to play softball, swim & I'm in a choir called Perpetual Music. I know that you have questions because I'll be 14 soon & I still have questions - so you can ask me anything you want & I promise to answer as best as I possibly can.

We're excited to bring this new feature to both the LiverLink and our Web site. Joe and Justine are happy to answer any questions that any AGS child, teen or young adult may have about life with AGS. They are here to offer support and suggestions based on their experiences with AGS. Questions may be submitted to alagille@teleport.com (or cchahn@worldnet.att.net), and can be responded to privately if necessary. Both Joe and Justine look forward to hearing from you!



Meet..... Justine Sawaya