
The Duct

Bringing you news on PSC



PSC PARTNERS
SEEKING A CURE

research, education, and Support

PSC Partners Seeking a Cure Foundation Spring 2009

ON THE ROAD TO CHICAGO:

A MESSAGE FROM RICKY SAFER, PRESIDENT OF PSC PARTNERS

This is one of my favorite times of the year. Here in the Colorado mountains, we're enjoying the peak of our wonderful ski season, while in Denver we're already seeing some early signs of spring. In addition, the countdown to our 2009 conference for PSCers and caregivers has begun. Becky Long and I are busy putting the finishing touches on the weekend plans, and everything is shaping up well! We hope that you'll consider joining us in Chicago, May 1-3 for our fifth annual conference which is co-sponsored this year by Northwestern University Feinberg School of Medicine.

To learn all the details about our 2009 conference, please read the Winter 2009 edition of our newsletter by going to www.pscpartners.org and clicking on Newsletters. To access our conference agenda, go to <http://www.pscpartners.org/2009ConfAgenda.pdf> and to access the registration form, go to <http://www.pscpartners.org/2009ConfRegForm.pdf>.

If you've never attended one of our conferences and you're trying to decide if you're emotionally ready for this intense weekend, I can honestly promise you that our conference weekends are an uplifting experience for everyone involved.

Before our first conference, I, too, felt ambivalent. I wanted to learn about the newest information on PSC and I especially wanted to meet other PSCers, yet I feared that the conference could end up being

2009 Conference information on the Web

The agenda and conference registration form are on the web at these sites:

<http://www.pscpartners.org/2009ConfAgenda.pdf>

and

<http://www.pscpartners.org/2009ConfRegForm.pdf>

two depressing days out of my life. Although it may sound counterintuitive, these conferences filled with so much medical information and myriad personal conversations with other PSCers and caregivers always end up being a positive force in the lives of all the attendees. Medical knowledge can be powerful, and the strong bonds that we form with each other keep up my spirits all year long. If you have any questions or concerns about the conference, feel free to write to me at contactus@pscpartners.org or to Becky at beckylong98@aol.com . If you're planning on attending, be sure to make your hotel reservations as soon as possible while rooms are still available at our special rate, and send back your registration form before our April deadline.

We are still looking for additional sponsors for our conference, but I'd like to thank our present sponsors:

Gold level sponsors: Mobile Doctors (Dike and Rilee Ajiri)
Illinois Tool Works

Silver level sponsors: Ken and Patty Shepherd

Their support is greatly appreciated!

Fundraising Challenge

Even if you aren't able to join us at the conference this year, please think about asking your family, friends and colleagues to donate to our Itching for a Cure/Road to Chicago fundraising challenge. This year, we have a terrific challenge that we need to meet. An anonymous donor will match dollar for dollar all donations that we receive from now until the conference up to the amount of \$45,000. Please help us meet this generous challenge. Every penny sent into the Itching for a Cure/Road to Chicago fundraiser will be used directly for PSC research. Please do your part to help us find that cure for PSC that we all dream about...Every donation, large or small, moves us closer to our goal. For details on how you can join this fundraiser, please go to <http://www.pscpartners.org/RtC.htm>

Grant Program Progress

Right now, our Medical Advisory Board, headed by Dave Rhodes, is hard at work reviewing fourteen excellent grant proposals that we have received from PSC researchers worldwide. Unfortunately, we don't have the money to fund all of these proposals right now, but your contributions to Itching for a Cure/Road to Chicago will help us fund as many requests as possible now and in the future. We are so happy to see how many valuable PSC research projects are being proposed. It's up to us as a group to provide as much funding as possible! At our 2009 conference, Dave Rhodes will announce which grant proposals have been chosen by PSC Partners Seeking a Cure. Remember our motto: "Together in the fight, whatever it takes!" and join us in this fundraiser.

PSC Symposiums

I'm glad to see that PSC research is getting more attention lately. There are two important PSC symposiums already scheduled for 2009. EASL (European Association for the Study of the Liver) is holding a three day symposium on PSC June 21-23 in Oslo, Norway, directed by Drs. Boberg, Karlsen,

and Schrupf. On October 3, PSC Partners Seeking a Cure in conjunction with the University of Colorado Health Sciences Center will hold a symposium in Denver on “The PSC-IBD Link: New Insights and Future Directions,” directed by Dr. Greg Everson and Dr. Sean Colgan. Both of these conferences will encourage PSC researchers and hepatologists to dialogue various issues relating to PSC. In our future newsletter editions, we’ll give you updates about new information presented at these symposiums.

Happy Anniversary

This January marked the beginning of the fifth year of PSC Partners Seeking a Cure’s existence, and the tenth anniversary of the Yahoo online group. Thank you to the online group moderators Tiffany, Johnathan and Arne for building that group up to its present total of 1649 members! PSC Partners Seeking a Cure has come a long way in our first four years, and I’d like to thank all of you who have helped us in various ways. The list is too long for me to mention everyone, but I’d like to give a very special thank you to our guru David Rhodes, who keeps us all informed about every detail of our disease. The PSC Literature site that David and his wife Judy have created and which they maintain is the only site of its kind. The database now contains a total of 104,000 abstracts and an associated library of about 58,000 pdf files! When I look back at our first four years, I am so proud of our early successes: four well-attended conferences for PSCers and caregivers, a free top quality online newsletter, research proposals that we are funding, a website that is being redesigned and expanded, four affiliated online support groups, etc. We have been able to accomplish all of this as a totally volunteer organization with an amazingly low administrative fee of 2.8 percent!!! Thank you to all our volunteers who have made this possible. Here’s to many more years of working together and finding that elusive cure!

Hoping to see you in the Windy City in May! Get your questions answered, join our compassionate PSC family, share some laughs, and learn some new coping skills.

The Duct’s Poll

Non- Prescription Itch Remedies: What works for you?

We wondered what you do about itching, what you use for relief. Let us know your tips to cope.

Respond below and we’ll compile the results and report back in the next issue.

Send your thoughts to: newsletter@pscpartners.org

Caregiver Corner

Speaking the Language of Caregiving: The Well Spouse Association

Finding someone you can talk to or email who is undergoing similar struggles as a caregiver is the idea behind “You Are Not Alone,” the comforting slogan of the Well Spouse Association, a non-profit group offering support and information to spouses of the chronically ill.

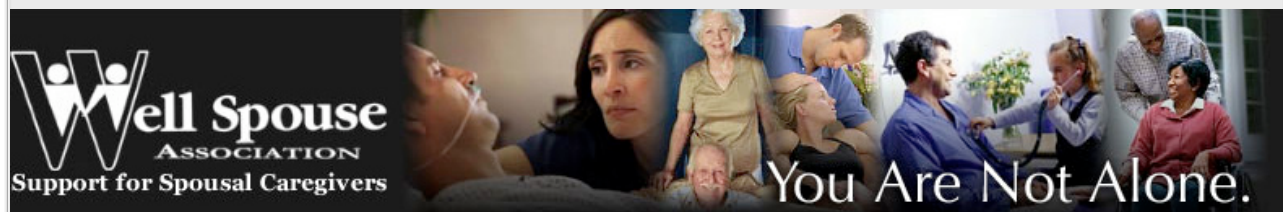
From the website (<http://www.wellspouse.org>):

“Well Spouse is a national, not for profit membership organization which gives support to wives, husbands, and partners of the chronically ill and/or disabled. Well Spouse support groups meet monthly. Here, our members can share their thoughts and feelings openly with others facing similar circumstances in a supportive, non-judgmental environment. WS support groups are also an excellent source for information on a wide-range of practical issues facing spousal caregivers.”

The easy to navigate site offers help in rebalancing life, invaluable information on the technicalities of caregiving, a landing zone of understanding about the experiences of helping an ill spouse, and support for developing a “new normal” even in the midst of a chronic health condition.

Joining the group costs \$25 for yearly individual membership. Membership in the organization offers support groups in many states, a quarterly newsletter, *Mainstay*, an online mentorship program, round robin letter writing groups of 5-7 for those who want to communicate via mail, respite weekends, on-line forums, and a one-day workshop on the Caregiver Journey, which is offered around the country. On-line forums include topics such as Help and Healing, Well Spouses Are People too!, Parenting and the Family, and Intimacy and Personal Issues.

The Well Spouse Association offers an annual conference, usually held in October, that is announced on the website.



Respond to: newsletter@pscpartners.org. Tell us what you'd like to read about regarding caregivers in future newsletters. If you have particular tips or a special point of view, why not share with others?

MY WORDS, MY STORY

Common Goals: Hoops 4 Healing and PSC Partners

Chuck Cooper, of Wooster Ohio, talked about Hoops 4 Healing at the 2008 PSC Partners Conference.

In May of 2008 I attended my first PSC Partners Conference in Jacksonville, Florida. To say I was amazed and impressed with what this organization has accomplished would be a huge understatement. I was also struck by how closely related the goals of PSC Partners and Hoops 4 Healing are and how much we have in common.

Hoops 4 Healing began back in 1998 with an almost improbable occurrence. Four men, all current or former basketball coaches, all living and working within 30 miles of each other, and all totally unaware of each other's conditions, formed a unique bond when we were all diagnosed with PSC. As we studied and learned more about the disease we realized that the chances of all of us being diagnosed with such a rare disease within a few years of each other were almost unbelievable.

As we compared our situations and learned more about our disease we decided we needed to organize ourselves and find a way to get in the fight against

PSC. The Hoops 4 Healing Foundation was formed with a simple goal in mind. That goal is to raise money to help find a cure for liver disease while increasing public awareness of the need for organ donation.

Because of the high profile nature of our jobs, our stories became public and we have had the opportunity to put more of a spotlight on the importance of each of these causes. Through a feature on the Today Show and an interview with the *Chicago Tribune* following Walter Peyton's announcement that he had PSC, we've had the opportunity to reach people in our area and around the country, to share our stories and help spread awareness of the desperate need for organ donation.



Each year we have attempted to expand our efforts to include new ideas to raise awareness. Since 1999 one of our most successful events has been the Hoops 4 Healing Basketball Classic in Canton, Ohio. This event includes both high school and college games and has featured local teams against great competition from around the country.

In 2002 we expanded our efforts to include a Golf Benefit, Dinner and Auction. Organized by NBA official and H4H board member Joe DeRosa and his wife Patti, this event, which featured an outstanding golf course, a great dinner, and the opportunity to bid on many special sports-related auction items generated approximately \$25,000.

In 2008 we were able to expand the basketball classic and widen our target group, by including a separate event for women's teams at the College of Wooster in Wooster, Ohio.

These and other fundraising events have raised close to

\$175,000.00 and put that money to use in the fight against liver disease. Our list of donations has included \$10,000 to the Cleveland Clinic, \$5,000 to Lifebanc, the organ procurement organization for Northeast Ohio, and \$15,000.00 to Mayo Clinic for research and development of liver disease cures.

I want to take just a moment and briefly tell you a little about our original four members because it's a good illustration of the depth and breadth of PSC. Bruce Brown is a former basketball coach at Bowling Green State University and is currently the athletic director at Lake HS in Uniontown, Ohio. Bruce was diagnosed with Crohn's disease in 1968 and with PSC in 1995. He is presently asymptomatic and continues to monitor the disease through his doctors. If Bruce walked into your living room you would think he was right off the beach and perfectly healthy. He is very definitely holding his own against the disease.

I'm next in line. After coaching basketball at the high school and college level, I became the athletic director at Wooster HS in Wooster, Ohio. I was first diagnosed with Ulcerative Colitis in 1972. Beginning in 1993 I was hospitalized off and on for five years with what they thought was acute pancreatitis. I was finally

diagnosed with PSC in 1998. Fortunately for me my condition deteriorated quickly and I underwent a traditional liver transplant in July of 1999. Since that time I have been very lucky to have a great support group and have remained comparatively healthy. On Christmas Eve of 2007 my doctors told me the PSC had returned and that I would need another transplant somewhere down the road.

Hal Smith is the next member of what we call "the liver club for men." Hal is the very successful head coach and athletic director at Malone College in Canton, Ohio. He is among the top 50 winningest coaches of all time and is highly respected throughout the college basketball world. He has spoken before the state legislature about organ donation and helped in passing a law making it easier to become a donor in Ohio by signing up on your drivers license.

Hal was diagnosed with PSC in 1991 and underwent a successful live donor transplant in April of 2000. About 2 years ago Hal was told that his PSC had returned and that he would need another liver transplant. About a year ago he found out that his kidneys are also failing and that he would also need a kidney transplant. Hal slowly moved up the waiting list until he was lucky enough to

receive a liver/kidney transplant in October of 2008. He was in the hospital for almost 2 months but is getting stronger everyday. He continues to be the most positive and upbeat person you could ever meet.

Our last member is Steve Loy. Steve was the head Basketball Coach at Walsh University in Canton, leading his teams to great success, posting a record of 254-130 in 12 seasons. In 1997, Steve was diagnosed with PSC. His condition remained stable until August 2003, when he was diagnosed with cholangiocarcinoma. After a valiant fight, Steve passed away in February 2004. Steve's death was a huge loss for us in so many ways, but it has also strengthened our resolve to do everything we can to educate as many people as possible about the importance of organ donation, and the fight against liver diseases. Steve's wife Karen has stepped into his role with H4H and is a valued member of our team.

As you can see, our lives have been affected by PSC and transplants, and as a result the focus of our organization is a little different than Partners Seeking a Cure. But make no mistake, we want to be a part of this partnership and contribute to the great work that is going on with this group.

BOTH SIDES OF THE COIN

David Spiro is a freelance writer from Rochester, NY. Diagnosed with PSC at age 26 in 1985, he received his “gift of life” of a liver transplant in 1997. Prior to being an organ procurement coordinator, David was a NYC paramedic with a 22-year career in emergency medical services. He owes anything he does to the courage of his donor family.

How often is it that a person can truly empathize with those to whom they provide a service? In health care, it is not often that a provider has the opportunity to view him- or herself on the other side of the hospital bed. It is even more rare for a patient to have the opportunity to be a part of the process that was instrumental in saving his or her life.

As a PSC survivor and transplant recipient, I’m in a category of people classified as “rare.” From 1999-2000, I worked as an organ procurement coordinator (OPC) with the organ procurement organization (OPO) that serves Rochester, NY, as well as other parts of western and central New York State. This was an experience that few transplant recipients have ever had: to work with donor families and donors on a very personal level.

For PSC patients, understanding the transplantation process can be an overwhelming experience. Fortunately, an entire team of transplant professionals is in place to assist them—from surgeons, to transplant coordinators, to transplant pharmacists.

This process takes place over a protracted period of time, (often years) from diagnosis of disease, to transplantation. For donor families, their experience with the donation/transplant process is measured in days, if not hours. An OPC is more often than not, the first person they will encounter along the way.

One of several responsibilities that OPCs have is to offer families of those who have been declared brain dead the option of donating their loved one’s organs for transplant (known as cadaveric donation).

Encountering families that have lost loved ones was not an unfamiliar experience for me. Prior to moving to Rochester, I worked as a New York City paramedic. One of the more difficult aspects of that job was informing families of the death of a loved one. Unfortunately, there were no options to offer a family under the circumstance of a sudden cardiac death that took place in the home.

While both situations meant dealing with tragedy, being able to offer a family something meaningful in relation to the death of their loved one is a very satisfying experience. This is what an OPC can provide: comfort in a time of sorrow.

There is no professional schooling to become an OPC. There is no degree issued in this field. Many OPC’s are usually nurses, with experience in some type of ICU or other critical care setting.

Many OPOs also use paramedics, since critical care training is a large part of our experience as well. New coordinators are paired with more experienced ones, and training comes on the job. They are gate-keepers to the organ procurement and transplantation network, and are responsible for:

- Initial evaluation of potential donors
- Insuring that brain death testing is carried out in a manner consistent with good practice guidelines
- Entering patients into the United Network for Organ Sharing (UNOS) national donor/recipient database, to match potential donors with listed recipients
- Public and professional education on organ donation

My own road to becoming an OPC began even before moving to Rochester. The transplant coordinator at Mt. Sinai Hospital in Manhattan (where I received my liver transplant) who was taking care of me opened up the possibility of pursuing this as a career option.

She knew my wife and I would be moving here, and through a series of professional connections, was able to recommend me to the director of the OPO in Rochester. I was interviewed not long after our move, and I found myself in this brave new world.

We worked in teams of two at our OPO; one coordinator responsible for monitoring and evaluating the patient in the ICU (or whichever critical care unit they were on), and one responsible for operating room coordination.

I mostly handled operating room coordination (as well as acting as a surgical assist during the organ recovery procedure), but on several occasions would be with my work partner when approaching families to obtain permission for donation. While the rate for obtaining donation in our OPO was very high, some families chose not to donate.

It was difficult as a recipient for me to rationalize this decision, but this is when the professional part of me would kick in and respect their wishes. I often felt cleft in two; the professional side of me understanding what needed to be done and why,

and the personal side of me struggling with the notion that the family who donated their sons' liver to me went through this same process.

What is most often misunderstood during this process is that the medical team that is taking care of the patient in the ICU is never involved in the actual consent process. This is not their role. Their focus is solely on the care of the patient, up to the declaration of brain death. When consent is obtained by the OPC for donation, maintenance of the patient is taken over by the coordinators (with help from the critical care team) in preparation for organ recovery.

If all this sounds clinical, it certainly can be. What was wonderful though was the interaction of OPCs with families of the patient after consent was given. I witnessed a sense of relief pass over many family members in the midst of their grief. As hard as it is to see a loved one die, there is a sense of hope that seems to transcend tragedy. I often heard donor family members speak of the notion that their loved one "lives on" through those who have received their organs.

I felt very privileged to have been a part of this process, though it only lasted a year. Psychologically, I found it too difficult to continue working with donors, especially in the operating room. As I found out, I was not alone. Through a colleague in New York City, I learned that two other OPCs that were transplant recipients also faced similar challenges, and were not able to continue working for their OPO. At the end of the day however, I was able to view the world of organ donation and transplantation from both sides of coin. It was an experience I can never forget.

April is Donate Life Month. Check it out: http://www.organdonor.gov/get_involved/donatelifemonth.htm

The Road to a Canadian PSC Partners

Contributed by Eve Jedrzejewska, Andre Altaras, and Mimi Altaras, proud parents and aunt of 5 year old Sara (diagnosed with celiac disease, PSC/autoimmune hepatitis overlap and ulcerative colitis) and yes, thankfully, she is doing well right now.

When a year and a half ago my then three and a half year old daughter was diagnosed with ulcerative colitis I thought it was the hardest thing I would face in relation to her health. But even as I struggled with the diagnosis, I learned to take comfort in the fact that there were medications that would likely control her condition and that despite all the challenges the illness was not life threatening.

Finding solace became harder when nine months later she was also diagnosed with autoimmune hepatitis and PSC overlap. Suddenly we were faced with a reasonably rare illness, with no known cause and cure, but more frighteningly no effective treatment. The internet was no help – all the statistics lead to a progression to liver transplant or death. Really not a lot of comfort for parents of a beautiful, effectively healthy-looking child. The complete lack of control and helplessness was not only scary but also infuriating. Within weeks I knew more about the disease than our friend, an anaesthesiologist at Sick Kids

Hospital in Toronto. At every doctor appointment I was asked if I was a doctor since I knew so much about her illness. NO, NO, NO, I wanted to yell – I am an accountant (and yes it became a joke between my husband and me and we took great pleasure in informing the astounded doctors of our profession), but more importantly we are parents who would like to be educated and empowered when it came to our child's illness.

All my attempts to learn about the illness yielded finding out about the PSC Partners conference in Jacksonville, Florida.

There was clearly no way I was not going. As my husband had work commitments, my sister-in-law offered to attend the conference with me. Even before we came to the conference I had an inkling of what I needed to do. But what we found at the conference exceeded our greatest expectations – the bond we found with the people was unbelievable – here were individuals in the same boat as

us – I felt like I made immediate friends and allies in my fight for my daughter's health. Here were also individuals, that like me, started completely grass roots out of the need to take control over this disease. And this is really how the road to the Canadian foundation took originally the idea in my heart and took root at this conference – the need to take control and not just be a silent helpless bystander in the process.

Luckily for me my profession has armed me with some skills to get moving on the practical items and the conference armed me with both Canadian and international friends and supporters in our cause. We formed a little “Canadian” team (Aubrey and Caroline, Rachel and Abe, Paula, Mimi, Andre and I) to tackle the set up of the charity – and we were/are well equipped – although small in number, we are big in heart and have some of the skills necessary to set up the foundation, and most importantly we have the perseverance.

To register a charity in Canada is a reasonably straight forward, albeit onerous process. The real challenge comes into play if you want the charity to be able to donate outside of Canada and to be able to direct funds to research directly (as opposed to other registered Canadian charities). This makes the application process a lot more complicated, and ultimately affects how quickly an approval of the registered status would be granted.

Given all the wonderful work being done by PSC Partners and that a lot of research is being carried on outside of Canada, the ability to donate outside of Canada seemed the ultimate path we wanted to end up on, however we quickly found out, through consultations with our lawyer, that this would take a lot of time (and of course more money) due to the more complicated application process and the rigorous approval process by Canada Revenue Agency for all charities.

We have two options: either go for the more complicated registration first, or set up a simpler charity (which would be faster) and then convert it to the more complicated one. As with all choices there are benefits and downsides to both -- the simpler charity would be up and running faster, but would be unable to donate outside of Canada -- and vice versa for the more complicated one. Although all these considerations have slowed us down, we remained undeterred.

So what have we done to date? We currently have a drafted (by our lawyer) preliminary organizing document for the charity and have to

vet it, and potentially make small amendments depending on which path of registration we undertake. We need to explore ways to get registered -- and hope that with an association to PSC Partners we can have a registration application that is more likely to be granted approval for a more complicated charity without delays.

We have had discussions with the main hepatologist treating Sara (our daughter), Dr Ling, a lovely man, who has agreed to help us in any way he can (and this is a great offer indeed since Sick Kids is a leading children's hospital not only in Canada but in all of North America and is involved in a number of research projects including the STOPSC initiative), including sitting on the Medical Advisory Board to the Canadian Charity.

We need to complete the registration package, including an objectives section for the charity. We need to recruit additional members onto the Medical Advisory Board. And no small feat, we have to think of ways to raise funds in addition to what we can contribute -- but of course that is a little further off.

I can honestly say that I wish we were moving faster -- but with all the turmoil in the markets and sometimes in our personal lives, we have encountered a slow down due to the distractions -- but that is all it is -- a slow down. We will continue to move forward on our path so that there is a better hope for a cure for all of us and our loved ones.

We want to see YOU at the 2009 Conference
Chicago, May 1-3
Agenda, registration and reservation form are at
<http://www.pscpartners.org>

Resources

Gastro Web Sites of Interest

There are a multitude of reputable web sites focusing on gastroenterology topics directed to patients.

Pharmaceutical companies, possibly your own gastroenterologist's site, national professional associations, university medical center departments--the list of resources is enormous.

This short review of professional gastroenterology sites--those catering primarily to physicians--is offered as a start for your individual research.

While PSC-ers are focused on the liver and hepatology topics, many of us also have related gastro issues and need information on them.

As with all web research, review what you find with your physician, and consider the credentials of the site presenter. Many pharmaceutical sites (not reviewed here) have useful information for patients, but may promote their products on the site.

The American College of Gastroenterology Patient Education page offers FAQs, information on GI health and disease, and colonoscopy. The group offers a free patient newsletter.

Web address is:

<http://www.gi.org/patients/>

The American Gastroenterology Association Patient Center is the AGA's helpful portal to a wealth of information on gastro diseases. There are tabs that focus on what a gastroenterologist does, general digestive conditions (PSC, is not mentioned, but IBD is), diet and medications, procedures, such as endoscopy, and web resources.

The site address is:

<http://www.gastro.org/wmspage.cfm?parmi=478>

The American College of Gastrointestinal Endoscopy offers a host of downloadable brochures on topics such as, colon cancer screening, polyps, ERCP, capsule endoscopy, and others.

The site address is:

<http://www.asge.org/patientinfoindex.aspx?id=1022>

The North American Society for Pediatric Gastroenterology,

Hepatology, and Nutrition has a Family Resources section focusing on diseases, treatments, tests and links to other organizations. PSC is not listed under diseases. Liver transplant is listed, but information is three years old. One section of the page allows families to find a pediatric gastroenterologist.

The web site address is:

<http://www.naspghan.org/wmspage.cfm?parmi=291>

We Support Organ Donation

One of our foundation goals is to increase organ donor awareness. We encourage U.S. readers to visit www.donatelife.net and click on their state.

This site gives a state-by-state guide to the organ donation process.

This would be a good place for our members to start thinking about how to help locally, if they are interested.

Donated organs and tissue are shared at the national level; the laws that govern donation vary from state to state.

Therefore, it is important for you to know what you can do to ensure your decision to be a donor is carried out.

WHAT IS ASCITES?

You've probably heard the term ascites (uh-SITE-ase) mentioned with frustration and a sense of dread from fellow PSC patients, especially those on the transplant waiting list. Ascites is a complication of significant chronic liver disease. Let's take a quick look at the condition.

If you have ascites you should already be under the care of a specialist and of course following the treatment plan he or she recommends. See the sidebar (page 13) for a listing of reliable web sites that can help you understand ascites.

Ascites is a build-up of fluid in the abdomen (the peritoneal cavity) and is most commonly seen in liver diseases, but it can also occur with additional serious problems, such as heart failure, certain cancers, and with certain kidney diseases, among others.

When the fluid accumulates, it can cause trouble with breathing, and in severe cases it can cause a lack of appetite. The fluid presses on internal organs, making them work harder. Patients report shortness of breath, feeling full, or having pain. In more advanced

cases of ascites the abdomen swells, or distends, abnormally.

Gastroenterologists classify ascites this way: Grade 1 is mild and ultrasound is the primary diagnostic tool, since belly swelling is not severe; Grade 2 can be noticed by bulging flanks and shifting dullness within (found during a physical examination by your doctor); Grade 3 is clearly visible because of the swelling belly and the fluid's obvious movement. At this stage a patient's daily life is affected.

Diagnostic tests often done when ascites is suspected include blood tests (CBC, metabolic profile, liver enzymes, coagulation) and possibly a paracentesis procedure, or capture of the belly fluid by a needle. The team can discern the cause of the ascites with these tests. A serum-ascites albumin gradient (SAAG) test can define whether the portal vein is involved in the development of ascites. Also useful in diagnosis are CT and MRIs, angiography, and laparoscopy to rule out other potential causes if liver disease is not thought to be the culprit.

Treatment for most liver-related ascites may include salt restriction, taking "water pills" or diuretics, water restriction, and weight loss. In addition alcohol consumption should be reduced, if not eliminated. NSAIDs are

often restricted as well. Common medications include furosemide (Lasix) and spironolactone (Aldactone), both diuretics.

Some patients with advanced ascites will need to have a shunt installed. A shunt is a continuous drain from the abdomen. There are three common shunts: portocaval, peritoneovenous, and the transjugular intrahepatic portosystemic shunt (TIPS).

These are effective at removing the fluid, but are generally a way station to liver transplant. For some patients with ascites the fluid build up is controlled by paracentesis, withdrawal of some of the fluid on an outpatient basis. This may be repeated regularly as the fluid will continue to build up after each procedure.

Again this is of limited value because it does not correct the problem and there can be significant complications. One complication of ascites can be bacterial peritonitis.

Become educated on this condition, learn the terminology, and follow the recommendations of your specialist. Ask questions; do your research. Ascites can be a result of liver disease, and while it can often be managed not cured, frequently a transplant is recommended because of the underlying problem.

Good Web Resources on Ascites

Wikipedia offers a comprehensive ascites overview:

<http://en.wikipedia.org/wiki/Ascites>

The National Institutes of Health site gives the basics:

<http://www.nlm.nih.gov/medlineplus/ency/article/000286.htm>

The reliable Merck Manual offers additional information:

<http://www.merck.com/mmhe/sec10/ch135/ch135e.html>

This article from the Cleveland Clinic goes into more detail:

<http://www.clevelandclinicmeded.com/medicalpubs/diseasemanagement/gastro/ascites/ascites.htm>

As with ALL web research, double-check your findings with your health professional.

Help us turn \$45,000 into \$90,000 for PSC research!!!

Please join us in our quest to find a cure by doing your part to help us meet an incredible fundraising challenge-to raise \$45,000 between now and our conference in Chicago May 1-3, 2009. We have an anonymous donor who has very graciously offered to match donations to The Road to Chicago fundraiser dollar for dollar up to \$45,000.

**Attention all 20 and 30-Something PSCers
(or those nearly in that range!) and Facebook Fans:**

If you haven't already, now is a great time to join the PSC Partners Facebook page! Get in touch with other PSC-ers your age to share concerns, ask questions and discuss what life is like when you're young and diagnosed with PSC.

If you've ever needed a place to complain about the fact that you need to nap when all your friends want to do is go out and party, share how you deal with drinking, ask others about issues dealing with starting a family, etc., here's the place for you!

To join, simply log onto www.Facebook.com and follow the prompts to set up a free account. It's simple to do and just takes minutes.

Once you have your account, just type in PSC Partners in the search box under Groups and you'll be taken to the page. It's just that easy! Hope to see and hear from you soon!



New Book on Cholestatic Liver Disease

Reviewed by David Rhodes

We would like to draw attention of our readers to a new book, *Cholestatic Liver Disease*, by Keith D. Lindor and Jayant A. Talwalkar. The book includes a chapter on Primary Sclerosing Cholangitis:

The Book's Particulars:

Cholestatic Liver Disease by Keith D. Lindor and Jayant A. Talwalkar

Edition: illustrated; Published by Humana Press, 2008; ISBN 1588298388, 9781588298386; 188 pages

Over the past 2 decades, there has been a steady increase in knowledge associated with the clinical manifestations of cholestatic liver disease. In addition to a growing amount of information on the well-studied disorders of primary biliary cirrhosis and primary sclerosing cholangitis, there has been a rapid accumulation of data on less well-known but important topics such as overlap syndromes with autoimmune hepatitis, cholestatic variants of alcohol and viral disease, and cholestasis following liver transplantation. These emerging insights are complemented by further improvements of the diagnosis and management of cholestasis. Based on these clinical situations, "Cholestatic Liver Disease" provides useful information for individuals involved in the care of patients affected by aspects of cholestatic liver disease, and supplies scientific updates from leading experts which relate to the clinical evaluation and management of cholestatic liver disorders, useful for both learners and practitioners alike.

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primary sclerosing cholangitis,
cholangiocarcinoma, ursodeoxycholic acid

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autoimmune hepatitis, primary sclerosing cholangitis, overlap syndrome

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We were pleasantly surprised to see our PSC Literature web site cited on p. 73 of the book:

“The new Mayo Clinic Risk Score no longer requires a liver biopsy (available at <http://www.psc-literature.org/mrscalc.htm>) and can stratify patients into low, medium or high risk of disease progression.”

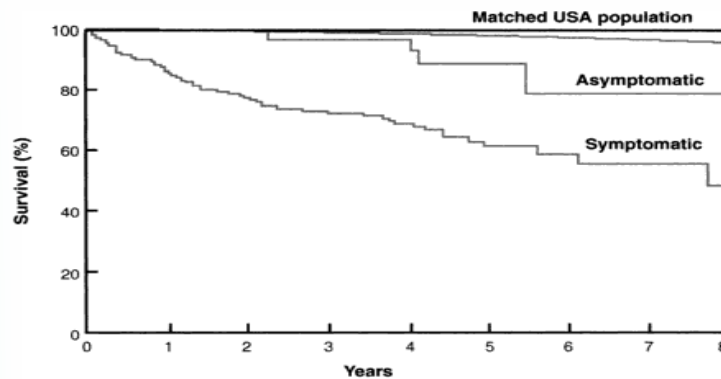


Fig. 3. The natural history of symptomatic and asymptomatic patients with PSC demonstrating decreased survival free of liver failure compared to age-matched controls (reproduced with permission from Ref. (37)).

histologic stage and age (12–15), with many including bilirubin as a marker of disease severity (12,13,15) and others including splenomegaly (13,14), hepatomegaly (14), alkaline phosphatase (14), or hemoglobin and IBD (12). The new Mayo Clinic Risk Score no longer requires a liver biopsy (available at <http://www.psc-literature.org/mrscalc.htm>) and can stratify patients into low, medium or high risk of disease progression (16). Others have suggested that the Child–Turcotte–Pugh (CTP) score is as good as the Mayo Clinic model in predicting outcomes in PSC patients (41), although the Mayo Risk Score is likely better at predicting prognosis in patients with early disease (42). The Model for End-stage Liver Disease (MELD), which is used to rank patients awaiting liver transplantation in the United States (43), has not been specifically validated in a large population of PSC patients.

Abstracts of Interest

by David Rhodes, Chair, Scientific/Medical Advisory Committee

Th17 Cells and Inflammatory Bowel Diseases

by David Rhodes

There have been remarkable developments in the understanding of autoimmune and inflammatory diseases in the last couple of years. The most significant developments have been the discovery that, in addition to the well-characterized CD4+ T cells called T helper cells type 1 and 2 (Th1 and Th2), two other CD4+ T cell types play a crucial role in regulating autoimmunity and inflammation. These two additional cell types are referred to as T regulatory cells (Tregs), which dampen or suppress

inflammation, and Th17 cells (so named because they produce interleukin-17) which promote inflammation. The factors which regulate the differentiation of these cell types was the subject of the recent “Hot Off the Press” page on the PSC Literature site, where we refer readers to a recent (free) poster from “Nature Reviews Immunology” depicting current understanding about these 4 cell types:

<http://www.psc-literature.org/HOTP.htm>

These discoveries mesh well with exciting developments in the identification of genes that affect susceptibility to inflammatory bowel diseases. It turns out that many of the newly discovered inflammatory bowel disease susceptibility genes are involved in the Th17 pathway. This was recently summarized by Abraham and Cho (2009):

[Annu. Rev. Med. 60: 97-110 \(2009\)](#)

[IL-23 and autoimmunity: new insights into the pathogenesis of inflammatory bowel disease.](#)

[Clara Abraham and Judy H. Cho](#)

The intestinal immune system has the challenge of maintaining both a state of tolerance toward intestinal antigens and the ability to combat pathogens. This balance is partially achieved by reciprocal regulation of proinflammatory, effector CD4⁺ T cells and tolerizing, suppressive regulatory T cells. Inflammatory bowel disease (IBD) comprises Crohn's disease (CD) and ulcerative colitis (UC). Genome-wide association studies have linked CD to a number of IL-23 pathway genes, notably IL23R (interleukin 23 receptor). Similar associations in IL-23 pathway genes have been observed in UC. IL23R is a key differentiation feature of CD4⁺ Th17 cells, effector cells that are critical in mediating antimicrobial defenses. However, IL-23 and Th17 cell dysregulation can lead to end-organ inflammation. The differentiation of inflammatory Th17 cells and suppressive CD4⁺ Treg subsets is reciprocally regulated by relative concentrations of TGFβ, with the concomitant presence of proinflammatory cytokines favoring Th17 differentiation. The identification of IL-23 pathway and Th17 expressed genes in IBD pathogenesis highlights the importance of the proper regulation of the IL-23/Th17 pathway in maintaining intestinal immune homeostasis.

Acronyms and Definitions

Allele: one member of a pair of different forms of a DNA sequence

CCR6: chemokine receptor 6

CD: Crohn's disease

CRP: C reactive protein

Genome-wide association (GWA) study: genome-wide comparison of common polymorphism frequencies between cases and controls

IBD: inflammatory bowel disease

IL23R: interleukin 23 receptor

Interleukin 12 (IL-12): consists of two subunits, p35 and p40, the latter of which is shared with IL-23

Interleukin 23 (IL-23): consists of two subunits, p19 and p40, the latter of which is shared with IL-12
JAK2: Janus-activated kinase
Polymorphism: a difference in DNA sequence between individuals
Proband: clinically affected individual through whom a family is found for genetic study
SNP: single nucleotide polymorphism
STAT3: signal transducer and activator of transcription
Th17: T helper subset 17
Treg: regulatory T cell
UC: ulcerative colitis

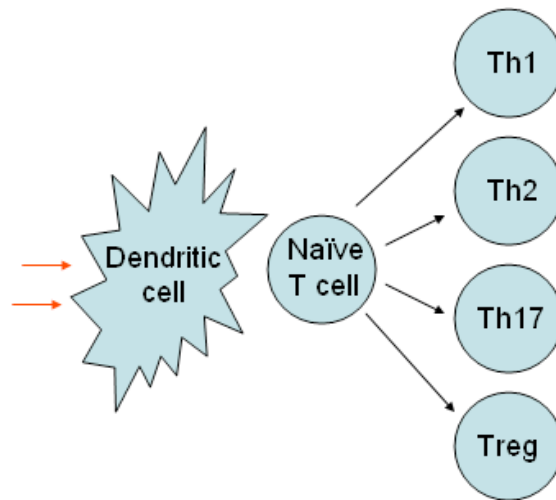


Fig. 1. Dendritic cells in the gut-associated lymphoid tissue, survey intestinal antigens (red arrows). They then interact with naïve CD4+ T cells to influence their development into different cell types: Th1, Th2, Th17, or regulatory T cells (Treg). The path of development of the T cells depends upon numerous cytokines and other factors [adapted from Abraham and Cho, 2009].

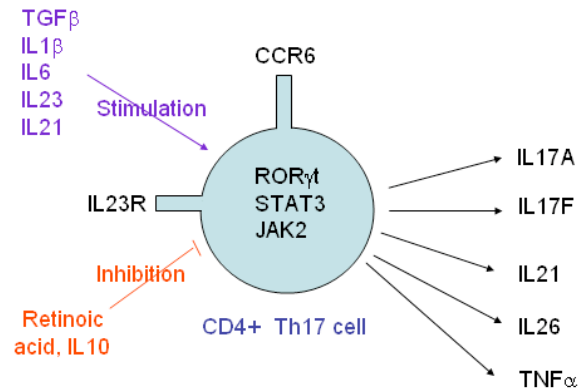


Fig. 2. Th17 cells are characterized by their expression of IL23R and CCR6 as surface proteins, and production of a number of pro-inflammatory cytokines such as IL17A, IL17F, IL21, and $TNF\alpha$. Th17 cell development is stimulated by cytokines such as IL6, IL23 and IL21, and inhibited by the vitamin A derivative, retinoic acid, and by IL10 (a product of Tregs). Genes involved in inflammatory bowel disease susceptibility include: IL23R, IL21, IL10, CCR6, STAT3, and JAK2 [adapted from Abraham and Cho, 2009].

Recent publications on susceptibility genes for ulcerative colitis are listed below. Particularly interesting are reports that IL-10 is a susceptibility gene for ulcerative colitis (note that IL-10 is produced by Tregs, which suppress Th17 cells), and that IL23R, STAT3, IL-21 and JAK-2 (all involved in Th17 cell development, as depicted in Figure 2 of Abraham and Cho (2009), above) are susceptibility genes for ulcerative colitis. In a commentary about the article by Anderson et al. (2009), Brandt (2009) states ... “In the present study, Anderson et al. provide further evidence that the Th17/IL23 pathway genes are associated with both IBD phenotypes: the IL23R ligand IL12B, JAK2, and STAT3 but not CCR6, were all significantly associated with UC at odds ratios very similar to that observed for CD.”

The genetic basis of ulcerative colitis is particularly relevant to PSC because over 70% of PSC patients have or will develop ulcerative colitis.

Recent publications on ulcerative colitis susceptibility genes

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Arisawa T, Tahara T, Shibata T, Nagasaka M, Nakamura M, Kamiya Y, Fujita H, Nakamura M, Yoshioka D, Arima Y, Okubo M, Hirata I, Nakano H 2008 The influence of polymorphisms of interleukin-17A and interleukin-17F genes on the susceptibility to ulcerative colitis. *J. Clin. Immunol.* 28: 44-49.

Brant SR 2009 Exposed: the genetic underpinnings of ulcerative colitis relative to Crohn's disease. *Gastroenterology* 136: 396-399.

Buning C, Schmidt HH, Molnar T, DE Jong DJ, Fiedler T, Buhner S, Sturm A, Baumgart DC, Nagy F, Lonovics J, Drenth JP, Landt O, Nickel R, Buttner J, Lochs H, Witt H 2007 Heterozygosity for IL23R p.Arg381Gln confers a protective effect not only against Crohn's disease but also ulcerative colitis. *Aliment. Pharmacol. Ther.* 26: 1025-1033.

Festen EA, Goyette P, Scott R, Annese V, Zhernakova A, Brant SR, Cho JH, Silverberg MS, Taylor KD, de Jong DJ, Stokkers PC, McGovern D, Palmieri O, Achkar JP, Xavier RJ, Duerr RH, Daly MJ, Wijmenga C, Weersma RK, Rioux JD 2009 Genetic variants in the region harbouring IL2/IL21 associated to ulcerative colitis. *Gut* Feb 6 [Epub ahead of print].

Franke A, Balschun T, Karlsen TH, Sventoraityte J, Nikolaus S, Mayr G, Domingues FS, Albrecht M, Nothnagel M, Ellinghaus D, Sina C, Onnie CM, Weersma RK, Stokkers PC, Wijmenga C, Gazouli M, Strachan D, McArdle WL, Vermeire S, Rutgeerts P, et al 2008 Sequence variants in IL10, ARPC2 and multiple other loci contribute to ulcerative colitis susceptibility. *Nat. Genet.* 40: 1319-1323.

Tedde A, Laura Putignano A, Bagnoli S, Congregati C, Milla M, Sorbi S, Genuardi M, Papi L 2008 Interleukin-10 promoter polymorphisms influence susceptibility to ulcerative colitis in a gender-specific manner. *Scand. J. Gastroenterol.* 43: 712-718.

Th17 Cells: Implications for New Therapies

By David Rhodes

So what do all these new discoveries concerning Th17 cells mean for new treatments of inflammatory bowel diseases (IBD)? At the outset, it is worth noting that Th17 cells are not only involved in IBD, but are also implicated in many extraintestinal manifestations of IBD, such as bone loss (Adamopoulos and Bowman, 2008), uveitis and scleritis (Amadi-Obi et al., 2007), psoriasis (Blauvelt, 2008), dry-eye syndrome (Chauhan et al, 2009), systemic lupus erythematosus (Garrett-Sinha et al., 2008), and spondylarthritides (Jandus et al., 2008), to name but a few examples. Therefore, treatments aimed at suppressing Th17 cells may have potential benefits not only in IBD but also extraintestinal manifestations of IBD. Moreover, these treatments are also of potential significance in transplant rejection (Chen and Wood, 2007)!

It is known that retinoic acid (a vitamin A derivative produced by dendritic cells in the gut associated lymphoid tissue), profoundly affects the balance between Th17 cells and Tregs (Mucida et al., 2009). Retinoic acid favors Treg production, and suppresses Th17 cell differentiation (Mucida et al., 2007; Kang et al., 2007).

The pro-inflammatory compound prostaglandin E2 appears to mediate its effects on inflammatory bowel disease via the IL-23 → Th17 pathway; thus, blocking prostaglandin E2 is an attractive target (Sheibanie et al., 2007; Chizzolini et al., 2008).

Because IL-23 stimulates Th17 cell development, antibodies to IL-23 would be expected to suppress inflammation. Indeed antibodies against the common subunit of IL-12 and IL-23 (p40) have been shown to be effective in treatment of both Crohn's disease (Sandborn et al., 2008), and the inflammatory skin disease, psoriasis (Krueger et al., 2007; Leonardi et al., 2008).

The metabolite of eicosapentaenoic acid (EPA), resolvin E1, has been shown to inhibit IL-23 and IL-6 production, raising the possibility that this may be one of the main mechanisms of the anti-inflammatory effects of fish oils (Haworth et al., 2008). Mudter and Neurath (2007) have suggested that blocking IL-6 is an important target for IBD treatment because IL-6 helps drive intestinal inflammation, by stimulating Th17 cells and suppressing Tregs. Franke et al. (2009) suggest that IL-10 (a product of Tregs) be re-evaluated as a potential therapy for ulcerative colitis in light of the discovery that IL-10 is a susceptibility gene in ulcerative colitis.

Blocking IL-21 would seem to be an intriguing target because IL-21 is not only produced by Th17 cells, but also stimulates Th17 cell production; thus, IL-21

tends to amplify its own production (Korn et al., 2007).

Recent patent applications give a sense of what some pharmaceutical companies may be considering for future medications and therapies for various autoimmune/inflammatory diseases. The following are three representative examples of recent patent applications and their disease targets. Note that **sclerosing cholangitis** is listed in all three.

1. This patent (<http://www.freepatentsonline.com/6579520.html>) seeks to block interleukin-17 and interleukin-17 receptor protein, with *potential* applications in the treatment of: systemic lupus erythematosus, rheumatoid arthritis, osteoarthritis, juvenile chronic arthritis, spondyloarthropathies, systemic sclerosis, idiopathic inflammatory myopathies, Sjögren's syndrome, systemic vasculitis, sarcoidosis, autoimmune hemolytic anemia, autoimmune thrombocytopenia, thyroiditis, diabetes mellitus, immune-mediated renal disease, demyelinating diseases of the central and peripheral nervous systems such as multiple sclerosis, idiopathic demyelinating polyneuropathy or Guillain-Barré syndrome, and chronic inflammatory demyelinating polyneuropathy, hepatobiliary diseases such as

infectious, autoimmune chronic active hepatitis, primary biliary cirrhosis, granulomatous hepatitis, and **sclerosing cholangitis**, inflammatory bowel disease, gluten-sensitive enteropathy, and Whipple's disease, autoimmune or immune-mediated skin diseases including bullous skin diseases, erythema multiforme and contact dermatitis, psoriasis, allergic diseases such as asthma, allergic rhinitis, atopic dermatitis, food hypersensitivity and urticaria, immunologic diseases of the lung such as eosinophilic pneumonia, idiopathic pulmonary fibrosis and hypersensitivity pneumonitis, transplantation associated diseases including graft rejection and graft-versus-host-disease.

2. This patent (<http://www.faqs.org/patents/app/20080299129>) seeks to develop antagonists to IL-17A, IL-17F, and IL-23P19, with *potential* applications in the treatment of: systemic lupus erythematosus, arthritis, psoriatic arthritis, rheumatoid arthritis, osteoarthritis, juvenile chronic arthritis, spondyloarthropathies, systemic sclerosis, idiopathic inflammatory myopathies, Sjogren's syndrome, systemic vasculitis, sarcoidosis, autoimmune hemolytic anemia, autoimmune thrombocytopenia, thyroiditis, diabetes mellitus, immune-mediated renal disease,

demyelinating diseases of the central and peripheral nervous systems such as multiple sclerosis, idiopathic demyelinating polyneuropathy or Guillain-Barre syndrome, and chronic inflammatory demyelinating polyneuropathy, hepatobiliary diseases such as infectious, autoimmune chronic active hepatitis, primary biliary cirrhosis, granulomatous hepatitis, and **sclerosing cholangitis**, inflammatory bowel disease, colitis, Crohn's disease gluten-sensitive enteropathy, and endotoxemia, autoimmune or immune-mediated skin diseases including bullous skin diseases, erythema multiforme and atopic and contact dermatitis, psoriasis, neutrophilic dermatoses, cystic fibrosis, allergic diseases such as asthma, allergic rhinitis, food hypersensitivity and urticaria, cystic fibrosis, immunologic diseases of the lung such as eosinophilic pneumonia, idiopathic pulmonary fibrosis, adult respiratory disease (ARD), acute respiratory distress syndrome (ARDS) and inflammatory lung injury such as asthma, chronic obstructive pulmonary disease (COPD), airway hyper-responsiveness, chronic bronchitis, allergic asthma and hypersensitivity pneumonitis, transplantation associated diseases including graft and organ rejection and graft-versus-host-disease, septic shock, multiple organ failure, cancer and angiogenesis.

3. This patent (<http://www.faqs.org/patents/app/20080219979>) seeks to develop antagonists to IL-17R (IL-17 Receptor A) with *potential* applications in the treatment of: inflammation, autoimmune disease, cartilage inflammation, and/or bone degradation, arthritis, rheumatoid arthritis, juvenile arthritis, juvenile rheumatoid arthritis, pauciarticular juvenile rheumatoid arthritis, polyarticular juvenile rheumatoid arthritis, systemic onset juvenile rheumatoid arthritis, juvenile ankylosing spondylitis, juvenile enteropathic arthritis, juvenile reactive arthritis, juvenile Reter's Syndrome, SEA

Syndrome (Seronegativity, Enthesopathy, Arthropathy Syndrome), juvenile dermatomyositis, juvenile psoriatic arthritis, juvenile scleroderma, juvenile systemic lupus erythematosus, juvenile vasculitis, pauciarticular rheumatoid arthritis, polyarticular rheumatoid arthritis, systemic onset rheumatoid arthritis, ankylosing spondylitis, enteropathic arthritis, reactive arthritis, Reter's Syndrome, SEA Syndrome (Seronegativity, Enthesopathy, Arthropathy Syndrome), dermatomyositis, psoriatic arthritis, scleroderma, vasculitis, myolitis, polymyolitis, dermatomyolitis, osteoarthritis, polyarteritis nodosa, Wegener's granulomatosis, arteritis, ploymyalgia rheumatica, sarcoidosis, scleroderma, sclerosis, primary biliary sclerosis, **sclerosing cholangitis**, Sjogren's syndrome, psoriasis, plaque psoriasis, guttate psoriasis, inverse psoriasis, pustular psoriasis, erythrodermic psoriasis, dermatitis, atopic dermatitis, atherosclerosis, lupus, Still's disease, Systemic Lupus Erythematosus (SLE), myasthenia gravis, inflammatory bowel disease (IBD), Crohn's disease, ulcerative colitis, celiac disease, multiple sclerosis (MS), asthma, COPD, Guillain-Barre disease, Type I diabetes mellitus, Graves' disease, Addison's disease, Raynaud's phenomenon, autoimmune hepatitis, and graft versus host disease (GVHD).

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Interested in Participating in a Mayo Study?

PROGRESS (PSC Resource Of Genetic Risk, Environment and Synergy Studies) website lists goals, participation requirements: http://mayoresearch.mayo.edu/mayo/research/lazaridis_lab/genomics_of_psc.cfm. The study asks for a small blood sample, and asks participants to fill out a questionnaire, all to examine genetic and environmental factors relating to PSC.

Our Photo Caption Contest: We Have a Winner!

Of course you remember that in the Fall issue this photo by Dr. Tom Karlsen of Oslo, a speaker at the May, 2008, conference in Jacksonville, inspired us to set up a contest for the best caption. When Dr. Karlsen set down his PSC Partners sport bottle and took in a glorious view, little did he know that the winner of the contest would be none other than his very good friend, Shelley Hussey, our favorite PSC Partners humorist.

Shelley's entry, "Peaking Tom!" was the winner, and her originality is now honored in the newsletter and fame will come her way, for sure. Congratulations, Shelley!





Itching for a Cure

Road to Chicago
PSC Partners Seeking a Cure Conference

NEW FUNDRAISING CHALLENGE: WE CAN DO IT!

Please do your part to help us meet this incredible fundraising challenge--to raise \$45,000 between now and our conference May 1-3, 2009. (See Ricky's Message on page 2.)

Our anonymous donor has very graciously offered to match donations to The Road to Chicago fundraiser dollar for dollar up to \$45,000. Help us turn \$45,000 into \$90,000 for PSC research!!! Every donation, no matter the amount, is welcome. For ideas on how you can participate in the Itching for a Cure Fundraising Challenge, go to the fundraising section of our website (<http://www.pscpartners.org/RtC.htm>). George Schill's animation is a happy addition to the effort.

Let's show our anonymous donor that we can live up to our motto: Together in the fight, whatever it takes! Let's move closer to finding that cure for PSC.

Be There or Be Square--Chicago in May: The 2009 Conference

You're invited for a weekend of learning, making and renewing friendships, inspiration, and fun in Chicago in May. Hobknob with the Northwestern docs, take a tour, attend a game, eat (!), play. It could even change your life.

Check out the agenda and conference registration form on the web at these sites:

<http://www.pscpartners.org/2009ConfAgenda.pdf>

and

<http://www.pscpartners.org/2009ConfRegForm.pdf>

Make Wyndham Hotel reservations, at special conference rates:

http://www.wyndham.com/groupeventsnew/orddt_pscpartners/main.wnt

The Glass Half Full

by Sandi Pearlman

As I'm sitting here writing this, I am mere hours away from my 33rd birthday. I suppose I could be out whooping it up, drowning the last of 32 in glasses of wine and beer held aloft by well-wishing friends and family.

Instead, I sit at home on my couch and find myself reflecting. To look at me, I look like an average girl, perhaps a little too thin, maybe in need of a haircut, but rather unremarkable all the same. No one would ever look at me and think, well, that girl there is fighting an incurable disease.

And yet, that's exactly what I am doing, what we are really doing (minus the girl for some of us!). We're fighters. We're given an impossible task: beat an incurable disease and live life without letting it destroy us, and, still, we march on.

I had a pre-interview with one of the counselors on my transplant team. For those of you who haven't yet had the pleasure, they ask you a lot of questions, try to gauge how emotionally

ready you'll be when and if the big day comes, freak you out about finances and planning and that kind of a thing.

In any case, my counselor (who would have a conniption if she read this and found I called her "counselor" rather than case worker or social worker) asked me what was the best thing that ever happened to me.

I responded that it probably hadn't happened yet. She frowned and asked me, well, what's the worst thing that ever happened to you. I said, well, it probably hasn't happened yet. Her response, I swear, was that she thinks I need therapy.

See, she couldn't understand why I wouldn't say PSC was the worst thing that ever happened to me. She couldn't grasp why it wasn't waiting there on the tip of my tongue like a cat after cheese and eager to drop from my lips. But the truth is, I find my answer hopeful that there's more to come, that PSC isn't the defining characteristic of my life.

Don't get me wrong, PSC sucks. I'm going to go out on a limb here and say that there isn't a single one of us who would choose to have the disease or keep the disease in the face of another positive option. But the fact is, in some ways PSC is both one of the best and one of the worst things in my life.

Before you all scream that the therapy idea was right, let me explain. The itching, nausea, RUQ pain, shortened life expectancy, etc., is more than a drag. It absolutely has to be worse than Chinese water torture.

But PSC also gives us an edge. We're not like the dude on the couch in the lounge somewhere thinking he has an unlimited lifetime to make choices and say sorry and make love and dream dreams. We know life is precious. It's ours for this day and hopefully for the next and, damn, if we're going to waste it.

We might be too tired to hit a party, can't go out for a beer with the pals or itch in inappropriate places at inconvenient times, but we know the value of the times we can go out, the friends we have and the blessed itch-free nanoseconds we're occasionally awarded.

I'm a glass half full kind of girl. In fact, some of my friends say relentlessly so. They insist if life threw at them what it's thrown at me, at us, that they'd curl up and die or spend all day crying in a corner and asking why me.

Well, the truth is, there are days when I want to curl up in a corner and whine why me and bawl my eyes out until I've lost

at least four pounds of water weight. But what's the use?

At the end of the day, unless you've got a better connection to God than I do, you're probably not going to get an answer. So, as 33 rapidly approaches, instead of wallowing in the have-nots of being sick and lacking energy and facing what could be years of poking and prodding and ERCPs and colonoscopies and the like, I find myself smiling.

I still may need therapy, but PSC has taught me to appreciate what is good. I know who my true friends are, I feel the warmth and the love as they and my family rally around me. I laugh as they jokingly refer to PSC as Pearlman, Sandi Cure. I think, all in all, I'm a lucky girl.

I may have a disease that requires a fight every single day of my life, I may be on a first name basis with more doctors than I can shake a stick at and have my own shelf in the pharmacy, but it also allows me to see all the things worth fighting for that might otherwise have gone unnoticed.

Thirty-three brings with it the first sign of wrinkles, a few gray hairs and a peace of mind that I can make a difference in my own life. I can raise money to fight PSC, to look for a cure. I can curl up and nap on a picnic blanket while my friends play Monopoly and drink wine.

I can be me without worrying what the world will think because I know every moment is precious and there's no guarantee that tomorrow will be there...and if it is and I'm itching and exhausted, well, I know I've survived it before and I'll most likely survive it again today.

Now on a completely unrelated matter, a plea to all of you 20 and 30 somethings reading this column.

I know most of you've heard us go on and on about the upcoming conference in Chicago on May 1-3, 2009, and, perhaps, about last year's conference in Jacksonville.

There's a reason for that. Quite simply, we want you to come! I'd say for a large number of us PSCers, especially when first diagnosed, we feel all alone out there in the world when it comes to all things PSC-related.

We hang on tight to our internet PSC Partners friends and rely on them for understanding, shared experiences, compassion, a good kick in the pants when needed; but, perhaps, we've never really met another PSCer face to face.

Well, the conference is your chance. It might seem scary, intimidating, depressing or just a waste of time; but I promise you, it's not.

Last year was my first conference. I went to it on the urging of my doctor (Thanks again, Dr. Harnois!) and was glad I did. I not only made fabulous friends, one of whom I talk to so often we almost need walkie-talkies, but just the experience of sitting down and talking to somebody who really, truly understands what it's like to have PSC is life-altering.

Suddenly, you're not the freak who can't stop scratching or can't make it through the day without a nap, you're one of many who can't stop scratching and can't get through the day without a nap. There's plenty of food, conversation and fun to go along with all the medical advice, research updates and statistics...and if that doesn't convince you...well, there's always the free pens and pill holders and such from our sponsors.

Just think, peace of mind, new friends and free gifts, I ask you, how can you beat that?

PSC Partners 2008 Financial Status

From our humble beginnings in 2005, PSC Partners has enjoyed financial growth year over year.

Income for 2008 was \$199,375, an increase of \$31,715 over 2007. The largest components of income are donations (\$20,410) and fundraising projects (\$125,270).

Total expenses were \$51,839 and included \$20,000 to the Morgan Foundation and \$6000 to the AASLD for 2 research awards. Administrative expenses are \$4275, only 2.8% of net income. All services are donated, the largest components of administrative expense

are D&O insurance of \$1150 and web service expenses.

Total net income for 2008 is \$147,536, compared to last year's annual total of \$143,684.

Total assets as of December 2008 are \$417,951, an increase of \$132,648 from Dec. 31, 2007.

As always, please direct any questions/ comments to Deborah Wentz, Treasurer at debs_3@charter.net.

Fact-O-Rama

Number of liver transplants in 2008: 5273

Number of liver transplants in 2007: 6495

Citation: <http://www.optn.org/latestData/rptData.asp>

National Average: MELD Score at Transplant, 2008

MELD Number transplanted nationally

<10	72
11-18	715
19-24	1708
25+	2204

Citation: <http://www.optn.org/atestData/rptData.asp>

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OCTOBER 2008-FEBRUARY 2009

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Chicago conference
PSC Partners Seeking a Cure

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Krogers Gift Cards
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Heather Lenihan

Sandi Pearlman
Fred Hussey

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IN-KIND DONATIONS

George Schill (for holiday card designs)
Jared Weinberger (for help with new logo design)
Scott Malat (for funding the new logo design)
Sue Safer (for designing new thank you cards and conference flyers)
Thank you for sharing your talents!

A BIG THANK YOU TO OUR NEWEST SPONSORS

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Gold level sponsors: Mobile Doctors (Rilee and Dike Ajiri)
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Thank you to all our donors for your support in funding our programs!

**Watch for the Big
Announcement:
First International PSC Day**

September 2009

Get Answers on PROGRESS . . .

Dr. Lazaridis will be speaking at the May Conference about the PROGRESS (PSC Resource Of Genetic Risk, Environment and Synergy Studies) project.

Erik Schlicht, program coordinator, will staff a booth at which participants can learn more and sign up to participate. See page 23.

RESEARCH STUDY OF PRIMARY SCLEROSING CHOLANGITIS AND VANCOMYCIN

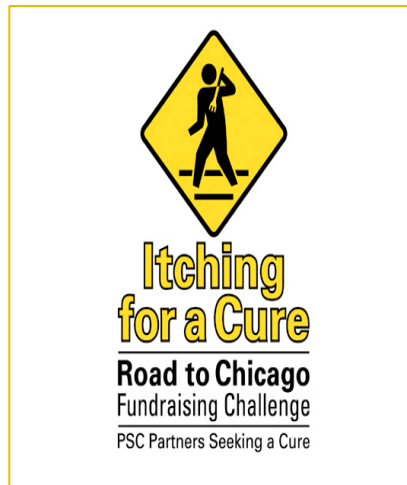
Massachusetts General Hospital is seeking volunteers, ages 18 years and older, with a known diagnosis of Primary Sclerosing Cholangitis (PSC) for a research study to see if a medication (vancomycin) improves liver disease symptoms, such as itchiness and fatigue and liver function tests (blood tests).

Participants should not be pregnant or breastfeeding, should not have any known allergy to vancomycin and should not have normal liver function tests.

Participation may last up to one year. Subjects will take vancomycin daily for 52 weeks and have follow up visits every six weeks.

Laboratory tests will be done at screening and every 12 weeks. A liver biopsy and MRCP (MRI of the liver) will be done at the beginning and at the end of the study. A flexible sigmoidoscopy will be done at the beginning of the study and at week 12. For more information, please contact Melissa at (617) 724-3238.

Our Latest Fund Matching Challenge: We Can Do It!



Note to Readers:

Articles in this newsletter have been written by persons without formal medical training. Therefore, the information in this newsletter is not intended nor implied to be a substitute for professional medical advice.

Please consult with your doctor before using any information presented here for treatment. Nothing contained in this newsletter is intended to be for medical diagnosis or treatment. The views and opinions expressed in the newsletter are not intended to endorse any product or procedure.



PSC Partners Seeking a Cure is a 501(c)3 nonprofit foundation that endeavors to find a cure for Primary Sclerosing Cholangitis.

The three-fold purpose of the **PSC Partners Seeking a Cure** foundation is to: raise funds for research on the causes and cures of PSC, promote PSC and organ donation awareness, and provide education and support to PSC patients and their families.

Ricky Safer is the principal contact person for the PSC Partners Seeking a Cure Foundation. Reach her at: contactus@pscpartners.org

Tax-deductible donations can be sent to:
PSC Partners Seeking a Cure,
5237 South Kenton Way,
Englewood, CO 80111 with a check made out to: PSC Partners Seeking a Cure.

Website

www.pscpartners.org

PSC Partners Seeking a Cure Board Members

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The Duct Newsletter

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