



Working together to provide research, education, and support for people affected by Primary Sclerosing Cholangitis

[www.pscpartners.org](http://www.pscpartners.org)

## PSC Partners Seeking a Cure Newsletter

### Vol. 2, Issue 2, May, 2006

Edited by David Rhodes and Ricky Safer

#### PSC Partners Seeking a Cure Board Members:

Dike Ajiri, Lee Bria, Elissa Deitch, Dr. Gregory Everson, Chris Klug, David Rhodes, Ricky Safer, and Deb Wentz

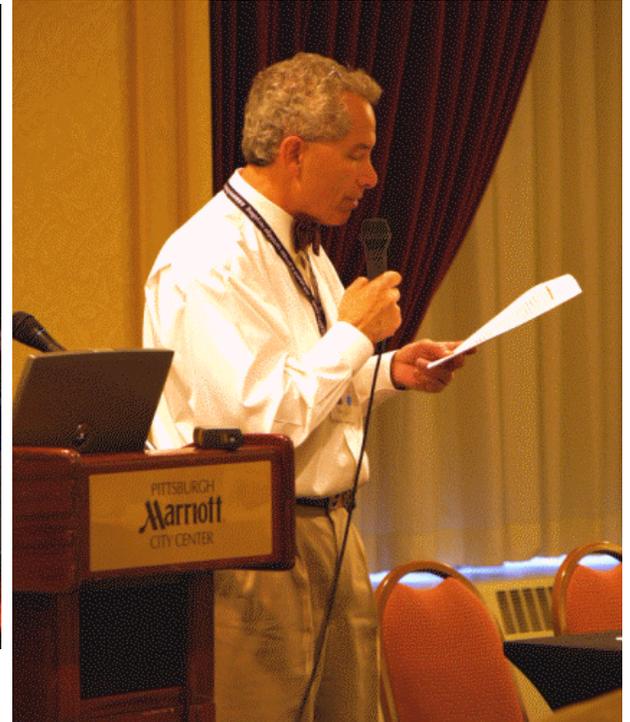
This issue of the newsletter is focused primarily on a summary of the **PSC Partners Seeking a Cure conference, held in Pittsburgh, PA, April 7-9, 2006.**

#### In This Issue:

- PSC - An Overview - Dr. Keith Lindor p. 2
- Role of the Endoscopist - Dr. Adam Slivka p. 2
- Itching in Cholestasis - Dr. Nora Bergasa p. 3
- Medical Management of PSC - Dr. Kapil Chopra p. 3
- Pampered in Pittsburgh p. 4-5
- Transplant Workup and Outcomes - Dr. Thomas Shaw-Stiffel p. 5
- Liver Transplant - Dr. Kusum Tom p. 6
- Organ Allocation - Nance Conney p. 6
- Attendee Comments from Pittsburgh Conference p. 7
- IBD in PSC - Dr. Leonard Baidoo p. 7
- Nutrition for PSC and IBD - Dr. Laura Matarese p. 8
- Update on PSC Research - David Rhodes p. 8
- Update on Dr. Chapman's Research - Ivor Sweigler p. 8
- The Morgan Foundation - Dr. Dennis Black p. 8
- Update on Donations to Partners Seeking a Cure p. 9
- Thank You p. 10
- 2006 Conference CD p. 10
- Autoimmune Liver Disease Conference p. 11
- Poetry Corner p. 12
- Special Thanks and Congratulations p. 12
- My First PSC Partners Seeking a Cure Conference p. 13
- Newsletter Article p. 14
- Cartoon (Presents for PSCers) p. 14
- Laughter is the Best Medicine p. 14
- Fundraising News p. 15
- Dr. Ulrika Broomé p. 16
- Making Donations to PSC Partners Seeking a Cure p. 16
- Additional Contact Information p. 16
- Submitting Newsletter Articles p. 16
- Note to Readers p. 16



Ivor Sweigler congratulates Mike Zaloudek when he arrived by bicycle at the conference on April 7. We were all in awe of Mike's three day, 270 mile, fundraising bike ride from Baltimore to Pittsburgh [photograph by Mark Stivers]



Master of ceremonies: Don Safer [photograph, and all subsequent photographs, by Arne Myrabo]



Dr. Keith Lindor

### Keynote Speaker, Dr. Keith Lindor, Mayo Clinic, Rochester, MN: **PSC - An Overview**

We were greatly honored to have Dr. Keith Lindor give the Keynote Address at the conference. Dr. Lindor defined PSC as a chronic cholestatic liver disease of unknown etiology, frequently associated with inflammatory bowel disease (IBD), and characterized by diffuse inflammation and fibrosis of the biliary tree, usually leading to biliary cirrhosis and portal hypertension. Whereas endoscopic retrograde cholangiography (ERCP) is most commonly used for diagnosis, magnetic resonance cholangiography (MRCP) is emerging as a non-invasive, cost-effective method that does not use radiation. Liver biopsy may help prognostically, and may assist in excluding other diseases. In the setting of cholestasis and IBD with a normal cholangiogram, liver biopsy may help in diagnosing small-duct PSC (affecting about 5% of PSC patients). Small-duct PSC can progress to classic PSC. PSC is a progressive disease with a 12-17 year median survival. A number of prognostic models have been developed; the new Mayo model uses age, bilirubin, AST, albumin and presence of variceal bleeding to assess risk of death. For management of cholestasis and pruritus, various medications are employed, including ursodeoxycholic acid, cholestyramine, naltrexone, and rifampin. Fat soluble vitamin deficiencies are common; especially vitamins A (40%), D (12%), and E (2%). Calcium and vitamin D supplements are important in preventing development of metabolic bone disease (especially osteoporosis). Hypercholesterolemia, steatorrhea, chronic pancreatitis and celiac disease may commonly occur. Dilation is preferable for management of accessible bile-duct strictures; long-term stents may cause problems. Specific complications of the disease include development of varices, and cholangiocarcinoma.

The latter may occur in 7-15% of patients with an incidence of 0.5% to 1% per year. CA19-9 has shown some promise as a tool for early detection of cholangiocarcinoma. Colon cancer occurs at a high rate in PSC patients with chronic ulcerative colitis. A number of medical therapies have been tested, but only silymarin, tacrolimus and high-dose ursodeoxycholic acid have shown possible benefit to date. In preliminary trials, high-dose ursodeoxycholic acid therapy is associated with improved liver biochemistry and portal inflammation, and indications of decreased disease progression, possibly translating to improved survival. However, long-term trials are still in progress. Ursodeoxycholic acid has shown some promise in decreasing risk of dysplasia in patients with chronic ulcerative colitis and PSC. Patients with autoimmune pancreatitis/cholangitis (characterized by elevated IgG4) may benefit from steroids. Liver transplantation remains the only successful therapy, with a survival rate of 90-97% after 1 year, and 85-88% after 5 years.



Dr. Adam Slivka

### **Role of the Endoscopist - Dr. Adam Slivka, UPMC**

Dr. Adam Slivka gave an excellent presentation on the diagnosis and management of PSC with emphasis on the role of the endoscopist. Endoscopic retrograde cholangiography (ERCP) is particularly important in 3 areas; diagnosis, managing complications (such as bile duct stones, cholangitis, and dominant bile duct strictures), and diagnosing cholangiocarcinoma. In diagnosis of PSC,

ERCP is the gold standard, and the key radiological features are irregularity and beading of intrahepatic and extrahepatic bile ducts. While MRCP is non-invasive, its accuracy is less than 100% and it offers no opportunity for therapeutic interventions and tissue sampling. ERCP offers opportunity for therapeutic interventions including stone extraction and placement of stents or dilation of dominant strictures. Dr. Slivka prefers balloon dilation and short-term (10-14 day) stenting, and tends to avoid long-term stenting. Dr. Slivka avoids sphincterotomy wherever possible, and notes that tissue sampling and liberal use of antibiotics are mandatory in these procedures. However, he concludes that there is no convincing data that the natural history of the disease is being altered by these interventions. Cholangiocarcinoma occurs in 15% of PSC patients and its diagnosis is extremely challenging. Tissue sampling (brush cytology, needle, and forceps) plays a key role in diagnosis of cholangiocarcinoma. While these techniques have relatively high specificity, they have low sensitivity. Serum tumor markers (CEA and CA19-9) together with brush cytology appear to improve the positive predictive value and accuracy of diagnosis of cholangiocarcinoma. Recent advances include combining these techniques with flow cytometry to determine DNA content of cells in bile duct brushings. Flow cytometry has shown promise in increasing sensitivity to 100% and specificity of cholangiocarcinoma diagnosis to 85%. Digital image analysis (to determine DNA ploidy) may also be a valuable adjunct to routine cytology. Dr. Slivka is particularly interested in using bile duct brushings to analyze genomic DNA for changes in a panel of specific microsatellite markers linked to tumor suppressor genes. Microsatellite allelic loss and mutations in a gene called "k-ras" seem to be highly predictive of malignancy. However, with increased sensitivity of diagnosis of cholangiocarcinoma comes a number of highly controversial issues, such as:

- how hard should we push for the diagnosis of cholangiocarcinoma, since it is usually a death sentence?
- should PSC patients be transplanted early as prophylaxis against cholangiocarcinoma?
- what is the role of living related donor transplantation in PSC with possible cholangiocarcinoma?



Dr. Nora Bergasa

### Itching in Cholestasis - Dr. Nora Bergasa, SUNY Downstate Medical Center

Dr. Nora Bergasa discussed pruritus (itch) in cholestasis. Pruritus is a common symptom of both primary sclerosing cholangitis (PSC) and primary biliary cirrhosis (PBC); both cholestatic liver diseases. It has long been thought that pruritus may result from impaired secretion of bile (i.e. accumulation of substances in the blood that would normally be excreted in the bile). However, the exact etiology of pruritus is unknown, and treatments remain unsatisfactory. It may become so severe as to be an indication for liver transplantation. Dr. Bergasa has had much experience in working with members of the primary biliary cirrhosis (PBC) organization to identify patterns in the perception of both pruritus and fatigue. She notes that in PBC the common sensations of itch are: feels like bugs crawling (35%), deep itch (29%), relieved by scratching (7%), and worsened by heat (41%). Seventy-three percent of PBC patients report that it interferes with sleep; 64% report that it is worse at night; and 19% that it is worse after meals. Sixty-seven percent of PBC patients state that doctors do not evaluate their itch in any way, and 28% report that they were not given any medication to treat pruritus. Cholestyramine (a bile sequestering agent), antihistamines, creams, antidepressants, ursodeoxycholic acid, and opiate antagonists were the most commonly prescribed medications, although a significant number of patients (11%) report that nothing has been helpful. Removal of pruritogens with plasmapheresis, partial external diversion, use of agents (such as phenobarbital, or rifampin) that induce hepatic enzymes, and use of ultraviolet light treatment of the skin, are sometimes effective. However, the absence of a consistent therapeutic plan suggests the need for further scientific investigation. Dr. Bergasa has noted that relief of pruritus does not appear to correlate with skin or plasma concentrations of bile acids, and that antihistamines are generally ineffective. Dr. Bergasa is particularly interested in the idea that opiates may be involved in pruritus, noting that opiate drugs (such as morphine) can often induce pruritus in humans, and scratching behavior in animals, and that opiate antagonists are effective treatments for opiate-induced pruritus. She has pioneered the use of the opiate antagonist naloxone as a potential treatment of pruritus, and has developed a unique apparatus (with a vibration transducer attached to the finger) to monitor and quantify scratching behavior (pruritic activity) in PBC patients (the scratch activity monitoring system (SAMS)). Dr. Bergasa has noted that in addition to opioids, a number of other neurotransmitters (such as serotonin, cannabinoid and gabaergic neurotransmitters) may also be involved in pruritus. She has initiated an International Society for the Study of Itch, and hopes that this will draw much needed scientific attention to this often neglected aspect of liver disease.



Dr. Kapil Chopra

### Medical Management of PSC - Dr. Kapil Chopra, Mayo Clinic, Phoenix

Dr. Kapil Chopra discussed medical management of PSC. He defined PSC as a progressive chronic cholestatic liver disease of unknown etiology that is commonly associated with chronic colitis. He noted that PSC usually leads to advanced liver disease and liver failure, and is an important indication for liver transplantation. Unfortunately, no effective medical therapy currently exists for PSC. PSC is often first suspected after an abnormality appears in a routine blood test evaluation. Most specialists use blood tests, cholangiography and liver biopsy to diagnose PSC. Many patients have no symptoms and may remain symptom-free for years. He emphasized several points mentioned earlier by Dr. Lindor, namely that the Mayo risk score can be used to estimate patient survival in PSC, and that the disease is associated with a number of complications, including decompensated cirrhosis, portal hypertension, and complications of chronic cholestasis (pruritus, fat-soluble vitamin deficiency, metabolic bone disease, hyperlipidemia, steatorrhea). Osteoporosis is common in advanced PSC. Osteopenia typically occurs in the lumbar spine, iliac crest and femur. Glucocorticoids used to treat IBD can aggravate osteoporosis. Patients are also prone to develop fractures after liver transplantation, although bone recovery can continue for up to 7 years after liver transplantation. Factors contributing to osteoporosis in liver disease include tobacco abuse, alcohol abuse, cholestasis, hypogonadism, and drugs. Factors contributing to osteoporosis after liver transplantation include: immobility, corticosteroids, malnutrition, reduced muscle mass/low body mass index, hormone fluctuation, and immunosuppression. While preliminary trials with high-dose ursodeoxycholic acid suggest improvement of liver biochemistry and reduced progression of the disease, further trials are needed to assess effects on long-term disease outcomes. Dr. Chopra gave detailed information concerning the ongoing multi-center, placebo controlled, randomized trial of high dose ursodeoxycholic acid (28-30 mg/kg/day) in PSC. This study will be the largest ever conducted in PSC and the follow-up will be the most extensive, providing an invaluable resource for studying the natural history of this disease. As part of this study, collection of serum, cells for extraction of DNA, bile, and tissue from the liver and colon will serve as a resource for future investigations. Primary end-points to be assessed include: histologic progression to cirrhosis, development of esophageal or gastric varices, need for liver transplantation, and survival. Secondary end-points include assessment of effects of high-dose ursodeoxycholic acid on liver biochemistries, histologic stage, cholangiographic features, Mayo risk score, and quality of life.

## PAMPERED IN PITTSBURGH

I'm just coming back down to earth after a rejuvenating, sometimes difficult, yet magical weekend. Thanks to Joanne Grieme and her committee, who have spent the last year planning tirelessly, the 2006 conference in Pittsburgh was a smashing success. We were thrilled to welcome 91 attendees from 24 states and two foreign countries. It was exciting to reconnect with many PSC Partners friends from last year and to welcome many eager new members into our group. It's hard to explain to non-PSCers why a weekend learning about our dreadful disease and sharing concerns with other PSCers and caregivers is not a depressing experience, but rather an uplifting weekend where I feel so much hope when we are together as a group.

As I look back on the weekend, here were a few highlights for me:

- I loved soaking up the positive energy that radiated throughout the room. Even though the bond that ties us together is a serious disease, our collective spirit is a very positive one.
- The slate of speakers was phenomenal, both in variety and overall quality. Dr. Keith Lindor's keynote address started out with a summary of PSC. Drs. Tom Shaw-Stiffel, Adam Slivka, Kapil Chopra, Leonard Baidoo and Kusum Tom offered us invaluable presentations combined with their compassionate attitudes. I was so impressed with the personal touch and caring attitude of the doctors at UPMC. I appreciated Dr. Bergasa's discussion of continuing research in her specialization on the itching problem that all of us PSCers dread.
- I was encouraged by Dave Rhodes' update on present PSC research and by Dr. Dennis Black's discussion of the future research plans of the Morgan Foundation. It is always enlightening to hear Ivor Sweigler's update on Dr. Roger Chapman's research in the UK.
- Two new adaptations to last year's agenda were a hit. The choice of six breakout sessions at the end of the day Saturday offered a wonderful change of pace, a lightening of the subject matter, and some very practical advice. Secondly, the Saturday night banquet, with Chris Klug as the keynote speaker, was the perfect way to end our marathon day. I think that everyone loved the chance to casually socialize with other PSCers and caregivers.
- When we broke into discussion groups on Sunday

(males with PSC/females with PSC/parents of PSCers/spouses of PSCers and other caregivers), the room came alive with heated discussions of our individual experiences with PSC and concerns about dealing with the disease. We all dealt with some difficult issues. I was glad to see that the male PSCers bonded, just as we women always seem to do naturally. Halfway through the roundtable discussions, Ivor came to ask me if he could join the female PSCers because "we were having more fun than his male PSCers." That's progress, boys!

- Thanks to Lee Bria, our always enthusiastic fundraising chairperson and cheerleader, our conference fundraising was a resounding success. Her idea of holding our first Virtual Walk was an incredible success, and will become an annual event. Thank you to everyone who participated. Start firing up now for next spring to challenge this year's leader, Mike Zaloudek!
- Cheering for Mike Zaloudek when he arrived by bicycle was another highlight. We were all in awe of his three day fundraising bike ride from Baltimore to Pittsburgh (270 miles!) I'd like Mike's secret on how to fight the PSC fatigue factor.
- I'm so encouraged with the way PSC Partners Seeking a Cure is expanding. On Sunday, I was touched to hear how many new members repeated the same message...that after this weekend, they no longer feel alone living with PSC and they finally feel some hope for the future.

We are extremely grateful to the following Corporate Partners who sponsored our 2006 conference: Astellas Pharma, American Liver Foundation-Western Pennsylvania Chapter, Axcan Scandipharm, Roche Pharmaceuticals, ConAgra Foods, UPMC-Thomas Starzl Transplantation Institute, and AAA Environmental.

A special thank you to Ali Lingerfelt-Tait for creating her new Andy Warhol-inspired design for our PSC all purpose notecards. They were a hot seller, along with the beautiful crafts of Alice Bennell, an energetic 17 year old PSCer from the UK. Her delicate beaded butterflies, knitted small purses and cross-stitch design were quickly purchased by the attendees. Thank you both for sharing your artistic talents to help our cause!

Thank you to the many members who offered practical suggestions on how to improve the foundation. We will try to follow through on many of these. Please keep all your input coming to us.

David Rhodes is already busy creating the CD of the conference presentations. When the CD is complete, he will mail a complimentary copy to each couple that attended the conference. Those who couldn't make it this year to the conference will be able to purchase the 2006 CD at a nominal fee (see page 10 of this issue). Thank you Dave!

Thank you to all our attendees who made this weekend such a success. You set the tone for all of us. We appreciate your support! To those who couldn't make it this year, you were greatly missed. We hope to see you at our 2007 conference in Denver.

Kudos again to Joanne Grieme for a spectacular weekend. You raised the bar for our future conferences.

I'm filled with conflicting emotions: sadness about PSC, pride for the courage and compassion of our members, and supreme disappointment that it will be another year before we convene again. I will miss all of you. I wish everyone an asymptomatic and healthy year.

Ricky Safer

Together in the fight, whatever it takes!



Dr. Thomas Shaw-Stiffel

### **Transplant Workup and Outcomes - Dr. Thomas Shaw-Stiffel, UPMC**

We were greatly honored to hear from Dr. Thomas Shaw-Stiffel concerning liver transplant workup and outcomes. Dr. Shaw-Stiffel initially defined liver failure. Liver failure is typically characterized by jaundice (yellowing of the skin and whites of the eyes), clotting problems

(easy bruising and bleeding), fluid build up in the ankles and stomach (ascites), and infection of the stomach fluid (peritonitis). This may also be accompanied by kidney failure [occasionally exacerbated or caused by various medications], internal gastrointestinal bleeding (varices), and/or mental confusion/coma (encephalopathy). Using an example of a case of a 28 year-old male with PSC, Dr. Shaw-Stiffel described how the patient progressed from initial diagnosis to liver failure, including the development of ascites, jaundice (elevated bilirubin), low serum albumin, and elevated creatinine (indicative of kidney failure). Dr. Shaw-Stiffel showed how the MELD score (using serum creatinine, total

bilirubin and INR (prothrombin time)) predicts 3-month mortality; a score which is used by UNOS to list patients for liver transplantation. Dr. Shaw-Stiffel showed how the aforementioned patient with PSC with serum creatinine of 1.9 mg/dL, total bilirubin of 4.2 mg/dL and INR of 1.2 had a MELD score of 20, implying a 50% chance of dying within 3 months. Dr. Shaw-Stiffel then proceeded to discuss how this patient was referred to the liver transplant team (hepatologist, social worker/psychiatrist, anesthesiologist, transplant surgeon), and evaluated. Typically such patients have blood work to determine blood type and HIV status, an echocardiogram, breathing tests, CT scan, Doppler ultrasound, and GI endoscopies as warranted. The eligibility of the candidate is determined by the transplant team, who then notifies UNOS for official "listing". The patient is closely followed by local physicians, and the transplant team is notified concerning any deterioration. The MELD score is recalculated at intervals. Typically the wait time is longest for O+ and A+ patients. Dr. Shaw-Stiffel noted that there is an organ donor crisis in the U.S., with 17,500 waiting for a liver transplant. However, only 4,500 will receive a cadaveric liver. The hepatitis C epidemic is the prime mover in this crisis. About 10% of patients will die awaiting a liver transplant. A problem specific to PSC is cholangiocarcinoma [the cancer is very difficult to diagnose and is often advanced when found]. Living donor liver transplantation (LDLT) offers a chance to remove the "bad liver". It is a valuable option because it provides less waiting time. Currently, over 2,500 living donor transplants have been performed world-wide. Because it is an elective operation, the transplant team is well prepared and rested. In LDLT the new organ is well studied, there is less concern about preservation, and generally the outcomes are similar or better than cadaveric liver transplantation. The downsides of LDLT are a 5-15% complication rate (mostly due to bile leaks), a 0.1% risk of donor death [there have been 2 deaths among 1,500 donors in the U.S.], and the financial burden to the donor (\$3,660 or more). Critical donor issues are voluntary commitment (no coercion), no financial or other incentives, extensive work-up to exclude intrinsic disease and to maximize donor safety, and a careful matching of liver size of the donor with the recipient. Potential donors undergo extensive evaluation, including phone interviews, blood typing, routine labs, interviews with psychiatrist and social worker, echocardiogram, ultrasound, MRCP and liver biopsy. Donor safety is the top priority. Dr. Shaw-Stiffel explained that liver regeneration occurs rapidly in both the donor and recipient following the operation. Dr. Shaw-Stiffel illustrated how the afore-mentioned PSC patient was successfully transplanted by LDLT, with his brother serving as donor. The donor was discharged after 6 days, and was back at work within 6 weeks, and the recipient was home within 3 weeks. Dr. Shaw-Stiffel noted that PSC can recur after liver transplantation, but there remain many unresolved questions concerning the frequency of recurrence, whether ursodiol helps, whether the recurrence is influenced by the type of immunosuppression used, and whether living donor recipients do better than cadaveric liver transplant recipients.



Dr. Kusum Tom

## Liver Transplant - Dr. Kusum Tom, Starzl Transplantation Institute

Dr. Kusum Tom focused on liver transplantation in PSC. She defined PSC as a chronic cholestatic liver disease, characterized by destruction of the biliary tract, often associated with inflammatory bowel disease, usually presenting in the third or

fourth decade of life, and with a male predominance. Clinical manifestations include pruritus, jaundice, abdominal pain and fatigue. As the disease progresses to cirrhosis, symptoms secondary to portal hypertension include: development of ascites, variceal bleeding, and hepatic encephalopathy. Cholangiocarcinoma is the most feared complication of PSC with an incidence of about 1.5% per year after PSC diagnosis is made. Cholangiocarcinoma has a survival of only 1 year, and is liver transplant contraindicated because of rapid recurrence and high mortality. Medical and endoscopic management of PSC was discussed earlier by Drs. Lindor and Chopra, and these aspects of PSC were therefore not reiterated by Dr. Tom. Rather, Dr. Tom noted that referral for liver transplantation is usually indicated when medical management fails, or due to poor quality of life (e.g. debilitating fatigue, intractable ascites, severe muscle wasting). Work-up for liver transplantation includes psycho-social, surgical and medical evaluation, endoscopy, cardiac work-up and radiological work-up. Listing for transplantation involves case presentation at a listing meeting, calculation of the MELD score (based on PT/INR, creatinine and bilirubin), and waiting for an appropriate cadaveric liver (wait times can range from months to years). Life after transplantation is usually associated with a marked improvement in quality, although there remain many concerns, especially in the areas of rejection, susceptibility to infection, and recurrence of primary disease. Post-transplant patients are usually on some form of immunosuppression for a lifetime, and infections are often controlled with prophylactic antibiotics. Living donor transplantation provides an alternative option to waiting for a cadaveric liver. The advantages of living donor transplants are: the surgery is scheduled, the organ to be transplanted is of known quality, the organ to be transplanted spends less time in cold ischemia, and surgery is not a race against time. The work-up for the living donor is similar to the workup for the transplant recipient and includes evaluation by the surgeon and hepatologist, CT scans, ultra sound scans, cardiac work-up, and liver biopsy. Surgery for the living donor transplants included 2 teams (donor and recipient). Typically for adult to adult live donor liver transplants, the right lobe is used (representing 60 to 70% of the liver by weight). The live donor program at the Starzl Transplantation Institute has conducted 122 adult to adult live donor liver transplants, of which 21% were for PSC. Patient and graft survival is 92%.

## Organ Allocation - How the Process Works - Nance Conney, Starzl Transplantation Institute

Nance Conney gave an overview of organ donation and allocation. She first discussed the history of transplantation, noting that the first transplant was a living-related kidney transplant in 1954. The first liver transplant was conducted by Dr. Starzl in Colorado in 1967. Cyclosporine, an immunosuppressant, was introduced in the 1970s (and finally FDA approved in 1983), and this markedly changed transplantation from research to treatment. The National Organ Transplant Act (NOTA) was passed in 1984, and in the same year the Organ Procurement Transplant Network (OPTN) was established; the United Network for Organ Sharing (UNOS) operates the OPTN. Nance Conney then discussed the organ donation process including key aspects of identification and referral, evaluation, consent, clinical management, organ allocation and distribution, recovery of organs, and follow-up. Evaluation usually involves a chart review and screening, physical assessment, a review of medical and social history, a determination of suitability, a determination of neurological status, and an assessment of organ anatomy and function; the latter determine acceptability. Various organs can be donated, including heart, lung, liver, kidney, pancreas and small bowel. Various tissues can be donated, including skin, bone, heart valves, connective tissue, fascia, eyes/corneas. Consent requires a medical examiner's or coroner's case to be cleared for donation, the death explained to the family, the donation options discussed with the family, and consent forms signed and witnessed. Clinical management involves tests for transmissible diseases, ABO blood typing, HLA typing, laboratory testing, and stabilization of hemodynamic functions. UNOS regulates all phases of procurement, placement and transplantation, and all potential recipients are registered. In the organ allocation and placement process, the key donor information needed is name, age, weight, height, ABO, donor hospital zip code, and available organs. Allocation of an organ depends on ABO compatibility, tissue type, medical urgency (e.g. MELD/PELD), height and weight range, time on the waiting list and distance from the transplant center. The MELD/PELD scores are used for liver transplant allocation for adults and pediatric patients respectively. All patients waiting for transplant are on the same national waiting list, which is blind to celebrity status, race, income and ethnicity. Yes, celebrities have been listed and transplanted quickly, but so have thousands of non-celebrities! Once an organ is donated, the organ is offered for transplant to the top of a UNOS generated list; the organ can either be accepted or rejected by the surgeon, and once accepted, recovery is coordinated with the transplant team and recipient. Organ recovery requires the coordination of recovery teams, an operative procedure, and organ preservation. Preservation times are limited to 12-18 hours for livers. Nance Conney urged liver transplant candidates to take care of themselves, to follow diet and exercise guidelines, and to make sure that they are always available for the call ... be prepared with transportation and packing!

## Attendee Comments from Pittsburgh Conference

*Here are some anonymous comments that we have taken either from e-mails post-conference or from our conference evaluation sheet. The attendees really capture the flavor of the weekend.*

I must admit that, by far, the greatest reward in making this trip was visiting with the wonderful members of our online PSC support group! The laughter and the love I experience when I am in the midst of these people is better treatment than anything any drug can provide!

I have been to several conferences, however this conference was the most organized, informative and worthwhile I have ever attended. We will be in Denver in 2007.

The conference was truly a healing experience. There was so much love, laughter and compassion in that group. I cannot express the inspiration and strength I got from being with all of you.

This group is the light left on in the darkness of my room.

You all keep my fears calm and my hopes high. The emotions of the weekend were strong, powerful and binding. The tears, cathartic. I have purpose and I have hope.

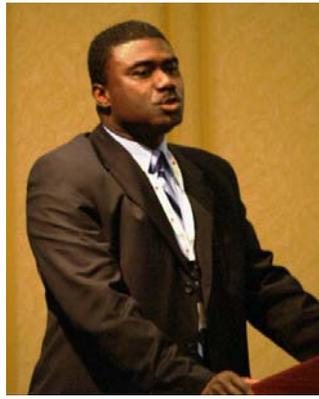
This was a very professionally done conference with a comfortable atmosphere. There was plenty of time to meet other people. Very impressive!

Before this weekend I felt so alone with this disease. What you have given me is renewed strength, encouragement, and a strong intestinal fortitude that I will beat this disease. For this, I am eternally thankful.

As a newcomer...thank you so much for the conference, the website, the welcome everyone gave me (as a new mom.) This group truly has given me hope for the future for my son and for all with PSC.

This is one of the most important events in our lives since our son was diagnosed. We now know that there are others too. The support, understanding and love from all of the group will help us through the challenging times ahead. We hope we will be able to help others too.

The conference was full of information, caring and partnership - the name of this group is so appropriate: PARTNERS Seeking a Cure.



Dr. Leonard Baidoo

### IBD in PSC - Dr. Leonard Baidoo, UPMC

Dr. Leonard Baidoo gave an excellent overview of inflammatory bowel disease (IBD), explaining how this is a chronic multi-system disease characterized by recurrent flares and periods of remission. There is currently no known cure, although numerous treatments can prevent complications. There are currently 1.5 million people in the U.S. affected by IBD, and the incidence appears to be increasing both in the U.S. and around the world. The current incidence rate is estimated to be 2.2 - 14.3 cases per 100,000 person years for ulcerative colitis (UC) and 3.1 - 14.6 cases per 100,000 person years for Crohn's disease (CD). IBD is often diagnosed at an early age (early 20's and 30's), and it spares no socioeconomic or ethnic group or race. The two types of IBD - CD and UC - may both be associated with extraintestinal manifestations. In UC, inflammation is limited exclusively to the colon (it may however be limited to the rectum or involve the whole colon) and usually presents with rectal bleeding, urgency and crampy abdominal pain. CD, however, occurs anywhere from the mouth to the anus. CD usually presents with bloody diarrhea, abdominal pain, fever and malaise, weight loss and loss of appetite, and failure to thrive. Extraintestinal manifestations of IBD can include erythema nodosum, pyoderma gangrenosum, uveitis and scleritis, sacroileitis, peripheral arthritis, and primary sclerosing cholangitis (PSC). It is estimated that ~ 2.4 to 7.5% of UC patients may develop PSC, and 1.4 to 5.5% of CD patients may develop PSC. The association between PSC and IBD was first described in 1985. Genetic, immunologic and environmental factors may be involved in both IBD and PSC. In the U.S., the percentage of PSC patients with IBD is about 75%, and of these, 87% have UC and 13% CD. In Japan, however, the percentage of PSC patients with IBD is much lower: 21-23%. The reason for this difference is unknown. Smoking may be one of the environmental factors influencing PSC, UC and CD. IBD can be diagnosed at any time during the course of PSC and *vice versa*, but in general it is diagnosed several years before PSC. PSC can develop even after total proctocolectomy for colitis. IBD can develop many years after liver transplantation for advanced PSC. The two diseases are so inter-linked that some people suggest that it be thought of as a variant of IBD ... PSC-IBD. PSC-IBD is often characterized by extensive colitis (pancolitis), rectal sparing, and backwash ileitis, a quiescent clinical course, a male predominance, and a higher risk of colorectal cancer. Factors influencing colorectal cancer incidence in IBD include severity, extent and duration of colitis, and the occurrence of PSC. The cumulative probability of colorectal cancer in UC alone is estimated to be about 10% after 25 years, but in PSC/UC it may be as high as 50% after 25 years. Early diagnosis can help prevent complications, plan treatment, and improve quality of life.



Dr. Laura Matarese

### Nutrition for PSC and IBD - Dr. Laura Matarese, UPMC

Dr. Laura Matarese discussed the importance of nutrition in PSC and IBD, including the basic principles of healthy eating, providing ideas for healthy snacks, providing suggestions for maintaining healthy weight, and giving a review of vitamin, mineral and electrolyte supplementation. She emphasized that good nutrition is essential for good health and energy, and to prevent nutritional deficiencies. Good nutrition does not mean that you have to sacrifice flavor and enjoyment! A healthy diet consists of: fruits, vegetables, whole grains, fat-free or low-fat milk and milk products, lean meats, poultry, fish, eggs, and nuts. A healthy diet is low in saturated fats, *trans* fats, cholesterol, salt (sodium), and added sugars. She suggests limiting simple carbohydrates (such as sugar) and including more complex carbohydrates (such as pasta, potato, breads, cereals, whole grains, and fruits and vegetables [as tolerated]) in the diet. She gave examples of healthy meals, and snacks, and suggested avoiding sugar alcohols (which can cause diarrhea), and alcohol, and limiting intake of caffeine (a GI stimulant which can cause calcium excretion). Dr. Matarese encouraged eating small meals spaced regularly throughout the day to minimize the urge to “snack”. Dr. Matarese discussed the roles of the duodenum, jejunum, ileum, and colon in absorbing different nutrients, and emphasized the importance of fat soluble vitamin supplements (vitamins A, D, E and K) to avoid fat soluble vitamin deficiencies in PSC, and the need for vitamin D and calcium supplements to prevent osteoporosis. Hydration is best maintained with fruit juice, low-fat milk, bottled water, low caffeine drinks, or special oral rehydration solutions during IBD flares.



David Rhodes

### Update on PSC Research - David Rhodes, PSC Partners Seeking a Cure Foundation

David Rhodes gave an update on research focused on bile transport proteins, the genes encoding them, the nuclear receptors that regulate their expression, and various medications that influence the activity of the nuclear receptors. He described some animal models of sclerosing cholangitis, including the *mdr2(-/-)* and *cftr(-/-)* mouse models. Recent results obtained with norursodeoxycholic acid (norUDCA) in the *mdr2(-/-)* mouse model look very promising; this novel bile acid seems to protect against the development of bile duct injury. The *cftr(-/-)* mice develop sclerosing cholangitis when given colitis. Docosahexaenic acid (DHA) protects these mice against bile duct injury, and is the rationale for the recent trial of DHA in PSC. Recent progress in inflammatory bowel disease genetics suggests that the MDR1 and the pregnane X receptor (PXR) genes may be important in ulcerative colitis.



Ivor Sweigler

### Update on Dr. Chapman’s Research - Ivor Sweigler, PSC Support (UK)

Ivor Sweigler gave an update on Dr. Roger Chapman’s recent results on high dose ursodeoxycholic acid (UDCA) [30 mg/kg/d] treatment of PSC. UDCA was well tolerated, led to improved liver biochemistry [i.e. reduced ALT, AST, alkaline phosphatase and GGT levels], and an improved Mayo risk score, suggesting improved survival. Ivor also gave an overview of Dr. Chapman’s proposed PSC research, including studies of the role of regulatory T cells in the pathogenesis of PSC, the role of atypical ANCA antibodies in the disease, the effects of UDCA on colonic dysplasia and cancer in PSC patients, and the additive role of antibiotics with UDCA in PSC. For example, UDCA plus rifampicin (rifampin) will be investigated in early stage PSC. Further details will be forthcoming in Ivor’s next issue of ‘PSC News’.



Dr. Dennis Black

### The Morgan Foundation - Dr. Dennis Black, Morgan Foundation

Dr. Dennis Black described the mission of the Morgan Foundation (The Musette and Allen Morgan, Jr. Foundation for the Study of Primary Sclerosing Cholangitis): to sponsor and facilitate both basic and clinical research to discover new treatments and ultimately a cure for primary sclerosing cholangitis. This foundation sponsored an NIH PSC Conference held in Bethesda, MD in September 2005; the summary of the conference will be published in *Hepatology* shortly. The Morgan Foundation has established a PSC registry (STOPSC) [Studies of Primary Sclerosing Cholangitis] which will include PSC, autoimmune hepatitis and overlap syndrome pediatric and adult patients, and will house a database of information and tissue and DNA samples, facilitating collaborative, hypothesis-driven, multicenter research on this rare disease. Its objectives are to identify risk factors, including genetic and environmental factors, for development of PSC, and to understand the mechanisms involved in the pathogenesis of the disease. It hopes to facilitate identification of genetic factors in the predilection of the disease, disease severity, and response to treatment. Candidate genes include HLA haplotypes, CFTR, MDR3, and NOD2, as well as inflammatory mediator gene polymorphisms, and liver disease modifier genes. The STOPSC registry will also help develop diagnostic tests/approaches that can diagnose the disease at an early stage, as well as surrogate markers for severity, progression and response to treatment. The registry will assist in clarifying the relationship between pediatric and adult PSC, the natural history of the disease, the relationship to allied diseases such as inflammatory bowel disease and autoimmune hepatitis, and risk factors and markers for the development of cholangiocarcinoma. It will further facilitate the evaluation and comparison of different therapies in multicenter controlled trials.

## UPDATE ON DONATIONS TO PSC PARTNERS SEEKING A CURE

<i>IN HONOR OF:</i>	<i>DONOR:</i>	<i>IN HONOR OF:</i>	<i>DONOR:</i>
Samantha Wente	Helen and Steven Timm	Joey Hatchett	Stan Smith
William F. Bria III	Rosemarie Bria-Levine		Kim and Cameron Beck
	Darcy Ward	Matha Marvin	Thomas Schulteiss
	Robert McColl	Mary Wells	Peggy Ann and Cyril Talbot
Bill Wise	S and S Cycle Inc.		
Josh Miletic and Joyce Garris	Adele and Mark Maxin	<i>IN MEMORY OF:</i>	<i>DONOR:</i>
	Mary and James Nalepa	Thomas A. Rogers	Elizabeth Ann Schmeltzer
Carolyn Beaudrot	Kathryn and M.S. Purcell		William Olmsted
Dike Ajiri	Natasha Moulton-Levy		Michael Kane
Mike Zaloudek	David Zaloudek	Dorothy Tenenbaum	Sima and Clifford Malat
	Arnold Jenkins	Stanley Breite	Shirley and Sol Schneider
	Anonymous		S.I. Raphael Limited, Inc.
	Ruth LaFontaine		Mary Ellen Rohon
	Broadway Services Inc.		Norma and Melvin Breite
	Leach Wallace Associates		Mina Seeman
	Comm-Tronics		Sally and Irving Breitowitz
Deb Johnson	Leora and Don Ackerman		Marie Zeichner
Flora Braunschweiger	Carol and Roy Gross		Anita Miller and Martin Field
Ricky Safer	Jan and Chris Karras		Alaire and Alex Mitchell
	Paula and Stan Ullman		Linda and Andy Kleinberg
	Patti and Mike Ross		Dorothy and Charles Samuel
Giving Back	Dana and Ray Miletic		Asta and Sidney Sperber
PSC Partners Seeking a Cure	Dianne and Paul Maranto		Rita Dadaian/Reese Spencer
	Arne Myrabo		Deborah and Fred Fishberg
	Traci Downs		Anne and Ronald Friedman
	Patty and Ken Shepherd		Gary Woods
	Deb and John Wentz		Birdie and Arnold Schulman
	Joanne Hatchett		Susan and Richard Birchman
	Dike Ajiri		Michael Samuel/Jane Martin
	Marco Fina		James Minter
	Marco Fina's parents and sister		Fern Platt
	B and B Wholesale Greens		Michele McDonald

**THANK YOU**

Thank you to our new platinum level sponsors:  
Patty and Ken Shepherd

Thank you to our new gold level sponsor:  
Rosemarie Bria-Levine

Thank you to our new silver level sponsors:  
Rilee and Dike Ajiri  
Broadway Services Inc.  
Joanne and Stephen Hatchett

Thank you to our new bronze level sponsors:  
David Zaloudek  
Kathryn and M.A. Purcell  
S and S Cycle Inc.  
Anonymous donation  
Kim and Cameron Beck

Thank you to our new copper level sponsors:  
Jan and Chris Karras  
Ruth LaFontaine  
Leach Wallace Associates  
Comm-Tronics  
Dianne and Paul Maranto  
Stan Smith

Thank you again to our corporate sponsors for our 2006 Conference:

Gold level: Astellas Pharma

Silver level: American Liver Foundation - Western Pennsylvania Chapter

Bronze level: Axcan Scandipharm

Copper level: ConAgra Foods  
UPMC-Thomas Starzl Transplantation Institute

Other sponsors: AAA Environmental



**2006 Conference CD**

The CD for the 2006 Conference is now available. Conference attendees will receive 1 free copy per household.

Copies of the 2006 Conference CD (\$10 each in the U.S./\$15 abroad; this includes shipping and handling) can be purchased from:

**PSC Partners Seeking a Cure**  
5237 So. Kenton Way  
Englewood, CO 80111

Please make checks out to: **PSC Partners Seeking a Cure**

Please include your name and mailing/shipping address when you place your order! Thank you!

Working together to provide research, education, and support for people affected by Primary Sclerosing Cholangitis

**CONFERENCE FOR PATIENTS AND CARE GIVERS**  
"TOGETHER IN THE FIGHT, WHATEVER IT TAKES"

**APRIL 7-9, 2006**  
**PITTSBURGH, PA**

Sponsored by:

Please locate and double-click on the file called "index.htm" on the CD. This file will link you to all of the content, including images and PowerPoint Presentations.

**Thomas E. Starzl Transplantation Institute**





# AUTOIMMUNE LIVER DISEASE

**Saturday, July 22, 2006**

**WASHINGTON INN • 495 Tenth Street • Oakland, CA • 10:00AM-1:00PM**

- **10:00AM**    **Overview of Autoimmunity and Emerging Therapies**    **Chris Bowlus, MD.**
- **10:15AM**    **Primary Biliary Cirrhosis**    **Marion Peters, MD.**
  - **Natural History/Complications**
- **10:45AM**    **Primary Sclerosing Cholangitis**    **Chris Bowlus, MD.**
  - **Natural History/Complications**
- **11:15AM**    **Autoimmune Hepatitis**    **Aijaz Ahmed, MD.**
- **11:45AM**    **Liver Transplantation for Autoimmune Liver Diseases**    **Stewart Cooper, MD.**
- **12:15PM**    **Research Update in Autoimmune Liver Diseases**    **Panel**

RESERVATIONS REQUIRED

NO CHARGE TO ATTEND

Name \_\_\_\_\_

Address \_\_\_\_\_ City \_\_\_\_\_ State \_\_\_\_\_ Zip \_\_\_\_\_

Email \_\_\_\_\_ Phone \_\_\_\_\_ Fax \_\_\_\_\_

PBC \_\_\_\_\_ PSC \_\_\_\_\_ AUTOIMMUNE \_\_\_\_\_ OTHER \_\_\_\_\_

**This program is suggested for patients and their families. A buffet lunch will be provided. Non-validated parking is available. Located 2 blocks from Oakland CitiCenter BART station.**

**FAX COMPLETED FORM TO 415/248-1066**  
**1/800-292-9099 x 14 • [www.liverfoundation.org/northernca](http://www.liverfoundation.org/northernca)**

### *"Do You Like Pina Coladas?"*

*We are just back from Fred's colonoscopy, and the whole experience made me wax poetic - I'm weird that way. Here's what the doctor said, more or less. (And I found it rather strange that she "sang" his diagnosis to him; even stranger that it was to the tune of "Do You Like Pina Coladas?")*

"Well you have ulcerative colitis,  
Got caught with your pants down,  
De-stress with daily yoga,  
On champagne we do frown.

You-also-have-a-pesky-parasite,  
Crawling all through your bowel,  
Take a second round of Flagyl,  
So he'll no longer prowl.

Here's an rx for a new med,  
Asacol is its name,  
It should end your diarrhea,  
Five months of ca-ca's insane!

(Now comes the musical bridge, followed by:)

What's up with this new heart murmur!?  
Go see a cardio doc -  
Echogram is what's called for,  
To check the tick in your tock.

If you like making love at midnight,  
And want to prolong your life,  
Stay away from caffeine/dairy,  
Make out a lot with your wife..."

(Anybody want the singing gastro's name and #, just email me.):)

Shelley Hussey,

Wife (07/28/78), of Fred, gang name: F-Dog, PSC 03/04, ADD (since birth), Bacterial Parasite 03/06, Pan UC (moderate case) 03/30/06, heart murmur 03/30/06 . . . Or as my brother just suggested after hearing all this: "Did Fred's warranty expire??"

### **Special Thanks and Congratulations**

Special thanks to Joanne and Stephen Grieme for all of their incredibly hard work in organizing the conference, and for making it such a huge success. Thank you also to Chris Klug for sharing his inspirational story. And congratulations to Chris and Missy on their engagement!

## **POETRY CORNER**

### **PITTSBURGH CONFERENCE**

We met in Pittsburgh to unite  
and bond in our common fight  
against the dreaded PSC  
and IBD (Crohn's and UC).

We saw vivid pictures  
of bile-duct strictures,  
of livers, and ascites .....  
not very pretty sights!

We learned about the role of ERCP,  
and other forms of cholangiography,  
about ursodiol, and its cousin "nor",  
the latest weapon in our war.

We heard of genetic studies,  
and made some buddies.  
We learned about the causes of itch,  
and made friendships rich.

We sought straight answers  
to our questions about cancers.  
We learned about clinical trials,  
and found time for smiles.

Between the many graphs  
we had some laughs.  
We experienced "Mike pride"  
For his amazing bike ride.

We aired our tribulations,  
and shared our hopes and aspirations.  
We confronted our fears,  
and shed some tears.

What a great conference!  
Thank you Joanne, Lee, Ricky and Don.

David Rhodes

Chris Klug, and Joanne and Stephen Grieme



## My First PSC Partners Seeking a Cure Conference

For those of you who don't know me, let me give you some background. My son Noah (8) was diagnosed with indeterminate colitis in March of 2005 and a PSC diagnosis followed in May after ERCP (and prior to that MRCP, liver biopsy...you know the drill). I have ALMOST been a member of this group for a year now. What a year it has been. How lucky am I to have found this amazing pocket of people in cyberspace? I just don't have enough words. There was no doubt that I had to attend the conference. There is only so long a person like me (very visual) can handle talking to people online all year without putting a name with a face. That is only one of the many reasons why it was important for me to attend.

Now for the confession, I was scared. I did not know what I would face. Too much information? Truths I could not face...yet? These people had all become family to me though, and I wanted to meet them, spend time with them...although not enough...and of course learn from them and the wonderful speakers who were scheduled. Well, I got all that and so much more. If you have been unable to attend, I encourage you to do so. This group is truly an amazing bunch, so accepting, so genuine, so sincere, so there for you. I am sad that any of us are in this situation that we find ourselves in, but after meeting you all, I know that we are going to be okay. We have each other and that IS something huge!

The first evening's reception was a blur for me as I really had not met anyone yet. I did meet Deb in VA on my way up to my room and I assumed she recognized me too...as I introduced myself to this "complete stranger." Deb is beautiful. So much pink in her cheeks...I think she looks better than me...and that is great!!! Her little Noah is a trip! Mike is just as I expected and I am sure totally, totally pleased that his wife is sitting next to him even if a pillow isn't fluffed.

Now for the talks: Wow! Joanne! What great speakers. What great everything. You are the bomb! You rock! How else can I say it? You just blew my socks off. Not to mention that you are mighty sweet too. I learned so much this first day...I took a deep breath and took it all in. I actually walked away feeling okay...maybe better. My husband heard some things that really freaked him out. (Like the cholangiocarcinoma survival times.) I thought I was going to have to scrape him up. I loved Dr. Slivka's pictures to help understand what is needed for good visualization of the small ducts.

After breaking for lunch, our table discovered the prior talk just created some more questions, so I took my list over to the transplant doctor from Starzl Transplant Institute, Dr. Kusum Tom and she graciously answered them. She was amazing. What an opportunity. I saw many doctors conversing before and after talks with the conference goers. I will just share what I was curious about. We were curious about what all has to match to be a good living donor? Dr. Tom indicated that they have transplanted across blood types. That was not even what I was expecting to hear. I just wanted to know that Noah and I being O+ was enough, that we did not need a tissue type to be the same, and we don't.

The evening dinner was a great time to relax after the knowledge we had obtained (exhale), and enjoy getting to

know each other. OH and the steak! To be honest, this was my favorite part. I love to chat...could you tell? Chris Klug was running around. We were in some great company. And can I just add here, what a truly NICE person he is. Nothing fake going on there. How refreshing! What an inspiration.

The next day consisted of mostly PSC Partners stuff. Ivor and David are amazing. We are truly a blessed bunch to have such great resources. They never get irritated with our humanness either. I am sure I have asked David the same question about something twice. He is always so pleasant. It was wonderful to meet him. Ivor proved to be everything I expected, very warm and kind. I only wish I could have spent more time with him.

This weekend for me was almost like a Marriage Encounter. All this stuff I have been doing for the last year, and talking about with the brick wall that wasn't listening (my husband) now was being put in a format he would assimilate. (He is a biologist. He did PCR stuff at Purdue on fish...this was right up his alley.) It was no longer Snoopy's teacher talking to him. I am going home not only less alone because he is now in my world, but less alone because I met people that I had never known in the group and met people I had known and I feel like I can finally breathe deeply now. We are all in the same boat, and we are all going to get through this. There will be bumps in the road, but I am so sure that we are going to make a difference.

Lee Bria is a powder keg of energy and that is what is needed for this fundraising endeavor, which was an amazing success. Wowzers! Mike Z...there is just nothing else to say and when you all see the numbers you will know why. Mike Z (we're not worthy...well, at least I'm not). Oh, and Mike and Aubrey... I am on Day 3 of butt desensitization on the bike front. Not so bad this time around. Ricky and Don Safer ... such a great, funny couple. Love the bow tie. Love the co-MCing. It was perfect! Just the comedy break needed at just the right time.

Melanie (MO) with all you are in the midst of, you are smiling, happy, blessing to everyone. I am so blessed to have met you online and LIVE! (and that you are girl!). Ali, oh sweet Ali, I always have a soft side for a girl from the south or at least with an accent. The cards turned out beautifully! You are truly a joy to be around. Jennifer, our blessed research advocate. Get the age lowered and I will help you out. You keep pushing girl. Keep that hubby's fingers tickling the ivory. Elizabeth (Liz), my sweet little caretaker, big hug! So glad to be getting to know you better. Dana, my dear Dana, Josh is so sweet. I am so glad you were there with me going through all of this. I don't know if I would have felt so good going if I did not think you would be there.

I could go on forever...literally...I know it is shocking...me going on.

You have to go next year. Start saving now to make it happen. It is worth every penny.

Deep peace and many blessings,

Maria Martens

## NEWSLETTER ARTICLE

By: Elissa Safer Deitch, outgoing legal counsel

I was sorry to have missed this year's annual PSC Partner's conference -- I heard it was wonderful. I also heard that many people at the conference were interested in knowing a bit about how the foundation is set up and how it makes its decisions. I thought I would just write a short summary describing all of this for those of you who are interested. If you want additional information, please always feel free to access our articles of organization at:

<http://www.sos.state.co.us/biz/ViewImage.do?jsessionid=0000911kcvTejIYBW-2vb7N-CN0:10e81rttv?masterFileId=20051002936&fileId=20051002936>

or contact Ricky Safer to obtain copies of any other corporate documents.

**ORGANIZATION:** We have been overwhelmed with the enormous involvement and support we've received for PSC Partners. However, initially, since we weren't sure whether we would be receiving this type of support, we organized PSC Partners to run very simply. PSC Partners is organized to be managed by its board of directors. The initial board was made up of the people who helped organize PSC Partners, and additional board members are elected by the existing board. PSC Partners also has members, but these members are non-voting members (ie they do not vote on decisions for the foundation). The current board members are: Dike Ajiri, Lee Bria, Elissa Deitch, Dr. Gregory Everson, Chris Klug, David Rhodes, Ricky Safer, and Deborah Wenthe.

**MAKING DECISIONS; PASSING RESOLUTIONS:** Under Colorado law (and pursuant to the foundation's bylaws), in order to make any decisions, or pass any resolutions, the Foundation can either have a meeting or obtain a written consent (without a meeting).

In order to make a decision or pass a resolution at a meeting: (a) a quorum must be present (either in person or by phone) at the board meetings -- since we have eight board members, this means that at least five board members must be present at the meeting. If we don't have a quorum present, board members can still meet and have discussions, but just can't pass any resolutions or approve decisions, and (b) once a quorum is present (ie the five members), any decision or resolution can be passed by a majority of the board members present (unless a higher percentage vote is required by law or the bylaws).

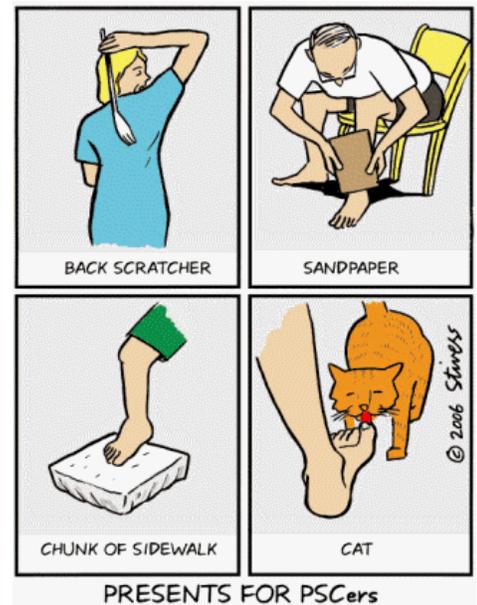
In order to make a decision or pass a resolution by written consent (without a meeting): (a) the Foundation must receive a signed written consent from each board member either approving or rejecting the resolution, and (b) at least a majority of the board members (or a higher percentage if required by law or the bylaws) must have approved the resolution.

**COMMITTEES:** The way that we have set up all our committees (and this includes the investment committee, the medical research committee, the fundraising committee, the treasury committee, etc.), is as follows:

(a) the board of directors gives the committees the authority to create guidelines and make presentations/suggestions to the full board of directors; and

(b) the full board of directors has to approve/vote on anything that is actually done (the committees don't have the authority to pass actions or resolutions without formal approval of the board of directors).

We're so excited to be working with all of you!



Cartoon by Mark Stivers

### Laughter is the Best Medicine

A new bile acid has recently been identified in tree ducks, swans and geese:

Kakiyama G, Iida T, Goto T, Mano N, Goto J, Nambara T, Hagey LR, Scheingart CD, Hofmann AF (2006) Identification of a novel bile acid in swans, tree ducks, and geese: 3alpha, 7alpha, 15alpha-trihydroxy-5beta-cholan-24-oic acid. *J. Lipid Res.* Apr 28 [Epub ahead of print].

We are looking for potential trade names and uses for this new bile acid (analogous to the trade names for ursodeoxycholic acid for the bile acid from bears that is used to treat cholestatic liver diseases; Actigall or ursodiol).

Suggestions for a name for this new bile acid include:

- AFLACgall
- quacktigall
- goosotriol

We suggest that it might be considered for the following conditions:

- duc(k)topenia
- other mal(I)a(r)dies of the bile duc(k)ts
- goostroenteritis

Dave Rhodes



Deb Wente (Treasurer), and Lee Bria (Fundraising Chair)

## Fundraising News

### Fundraising for PSC Partners Seeking a Cure is off to a great start for 2006

Thank you to all our members who virtually walked with us from our home towns to our conference in Pittsburgh! We had 52 members virtually walking. Together we raised an incredible \$44,493.29!!!! This is an awesome display of how working together we can help to beat PSC!!! This is fantastic and we are so proud of each and every shirt. The picture below shows some of the shirts that were on display at the conference.

Our winners for the most creative shirt were:

- 1st place.....Joyce Garris [Dana Miletic's mom]
- 2nd place.....Reggie and Jeff Belmont
- 3rd place.....Joanne Grieme

Most signatures were from Mike Zaloudek, who had over 300 signatures. Second place for signatures was Joanne Grieme with 151 signatures. Third place for signatures was Lee Bria with 90 signatures.

Most donations went to Mike Zaloudek, with an incredible total of \$19,017.65! Second place to Lee Bria with \$7,777! Third place to Joanne Hatchett with \$6,330! Each and every dollar raised for this fundraiser is appreciated! Every registered walker can be proud of their contribution. This walk shows the determined spirit behind our slogan of "together in the fight, whatever it takes". All of us working together will make the difference. Thanks to all!!!

### Silent Auction

Our silent auction had some fun items and brought in a total of \$807.39.

### Simple Gifts

A big thanks to Alice Bennell, for her wonderful contribution to PSC Partners Seeking a Cure. Her Simple Gifts raised a total of \$325 from her sales at home and sales at the conference. You are amazing Alice!

### Notecards by Ali

Our PSC Partners Seeking a Cure notecards designed by Ali Lingerfelt-Tait were once again a big seller. Her new design is terrific and the notecards can be ordered from our website. Everyone should have some handy. Thank you Ali!

### Wristbands

Wristbands are still available and can be ordered from our website. Having a fundraiser? The wristbands are great to hand out. With our website printed on the band, it is a great way to promote both PSC Partners Seeking a Cure and organ donation.

### AAA Update

All of us working together are making a difference just by recycling old cell phones and ink cartridges. This program isn't even a year old yet, and you can see that one ink cartridge and one cell phone at a time we are helping PSC Partners Seeking a Cure. Thanks to all of you who are recycling. For more information please contact Tim Wholey at: [timwholey@cox.net](mailto:timwholey@cox.net). Totals from August 2005 through April 2006 for recycling = \$636.57. Thanks to AAA Environmental for their generous donation of \$250 for our conference.

### Kroger Update

I'm looking for Kroger shoppers to help us with one of our best fundraisers! So far in 2005 and 2006 we have raised \$2,486.38!!! If you shop at Krogers you can help PSC Partners Seeking a Cure earn money from Krogers at no cost to you just by using a specially registered gift card. Please contact me at: [ldbria@comcast.net](mailto:ldbria@comcast.net).

A special thank you to Nichole Rowland whose creative fundraiser selling roses at a children's dance recital brought in \$622.35 for PSC Partners Seeking a Cure. Thank you also to Bud and Bettyanne Harlow for donating Beanie babies and greenery to this project.

Lee Bria



The 'Virtual Walk' T-shirts, with signatures, were displayed at the conference. Part of the display is shown here (photograph courtesy of Arne Myrabo).

All of us at PSC Partners Seeking a Cure really appreciate the hundreds of hours of pro bono legal work that Elissa Deitch has done for us. Without her, we wouldn't exist! Good luck with the new addition to your family, Elissa. We'll miss you!

The board of directors

We are thrilled to welcome these new PSC partners who have recently offered their much needed expertise:

- Sima Malat (mother of Scott Malat), whose office will prepare our year end tax returns at no charge.
- Larry Rich (father-in-law of Jason Drasner), who will share duties as legal counsel with: Nina (new member).

Thank you in advance for volunteering to help us with these important jobs.

### Dr. Ulrika Broomé

We were all very saddened to hear that Dr. Ulrika Broomé of Sweden passed away April 3, 2006. Dr. Broomé was one of the leading researchers on primary sclerosing cholangitis. She will be greatly missed by her patients and the PSC research community. For details of her impressive research papers on PSC and IBD, please see:

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

PSC Partners Seeking a Cure extend our deepest sympathies to her family, friends, colleagues and patients.

### Additional Contact Information

Ricky Safer is the principal contact person for our PSC Partners Seeking a Cure Foundation. She can be reached at:

[pscpartners@yahoo.com](mailto:pscpartners@yahoo.com)

### Submitting Newsletter Articles

If you would like to contribute an article to a future issue of this Newsletter, please e-mail it to David Rhodes:

[rhodesdavid@insightbb.com](mailto:rhodesdavid@insightbb.com)

## Making Donations to PSC Partners Seeking a Cure

Tax-deductible donations can be sent to:

**PSC Partners Seeking a Cure  
5237 So. Kenton Way  
Englewood, CO 80111**

with a check made out to:

**PSC Partners Seeking a Cure**

Alternatively, donations can be made on-line via PayPal (<https://www.paypal.com>) to [pscpartners@yahoo.com](mailto:pscpartners@yahoo.com)

Please include a note to indicate whom the donation is in honor and/or in memory of, and your return address.

We offer several levels of sponsorship

- Platinum level: \$10,000
- Gold level: \$5,000
- Silver level: \$2,500
- Bronze level: \$1,000
- Copper level: \$500

Thank you for your generosity!

One of our foundation goals is to increase organ donor awareness. We encourage U.S.A. readers to visit [www.donatelife.net](http://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....“While 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.”



**GiveLife**

### 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.