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Bringing you news on PSC

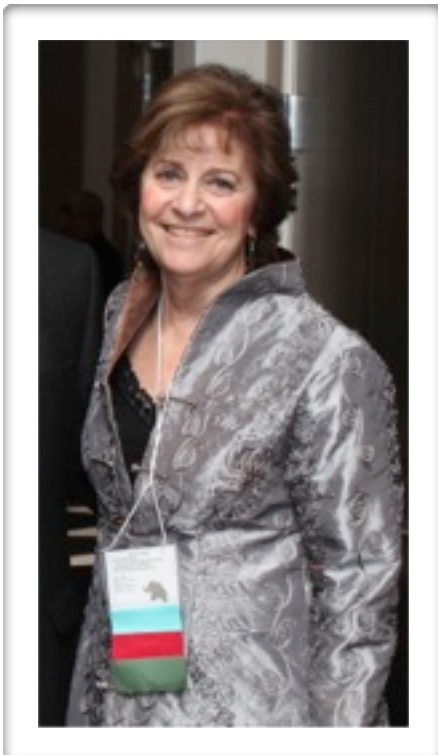


PSC PARTNERS
SEEKING A CURE

research, education, and Support

PSC Partners Seeking a Cure Foundation Summer 2012-I

That Special Indescribable Feeling: The 2012 Conference



by Ricky Safer

I've finally found the time to process this year's total conference experience. In particular, the variety of medical information, the sharing and advice from other PSCers and their caregivers, the sorrowful as well as the fun moments that we spend together as a group, and most of all, the ongoing camaraderie and support from everyone are "take aways" that last the whole year through. I try to explain to friends at home why spending a weekend with others living with our dreaded disease is always a magical experience, rather than the depressing event that they envision. Every year, I enjoy the months of planning the speakers and topics, the venue, the food, pre-conference activities, and all the other details. Truly, it is our unique community all working together that creates the special spirit and positive energy of our annual conference.

Where else do we have brilliant physicians volunteering their time and expertise to educate us about PSC while sharing their compassionate advice and promising information?

Where else can we be in a packed ballroom where everyone understands our unspoken problems, fears and hopes for the future? Where else can newcomers walk into a group of strangers, and within a short time, feel comfortable knowing that they are automatically part of

a welcoming community that is working together with a common goal? Our lives have been suddenly thrown together because of a terrible disease, yet when we are together, there is a magical energy that invigorates all of us. As our PSC community continues to grow each year, I worry that we will become so big that our community spirit will be broken, but

thanks to all of you, the positive, caring spirit is still alive and growing.



Our eighth annual conference for PSCers and caregivers took place May 4-6 in conjunction with the phenomenal physicians from the Mayo Clinic Rochester. In addition to thanking all our wonderful speakers from the Mayo Clinic, I'd like to give a special thank you to Dr. Keith Lindor, Dr. Kostas Lazaridis and Dr. Jay Talwalkar for planning this year's excellent dynamic agenda. We offered some

new topics this year, such as sleep and chronic illness, the MELD score, understanding PSC medications, and the mind-body connection. All the expert physicians and health professionals made it clear why the Mayo Clinic has earned its incredible reputation! The speakers did a remarkable job explaining complicated medical procedures and concepts at a level that our audience of laypeople could easily understand. In addition, the speakers made themselves available during the day for our attendees to speak with them one on one. We all learned the latest information on all aspects of life with PSC as well as the promises of future PSC research.

This year's attendance was our largest ever - 252 attendees! The participants came from 31 states within the United States and represented eight foreign countries. There were 21 Canadians, as well as attendees from the UK, the Netherlands, Sweden, Norway, Israel, Australia and South Africa. We appreciate the time and effort that it takes



each and every one of you to join us. PSC may be an unwanted disease, but when we face it together and work closely as a group, we end up feeling positive that we are no longer alone, and that as a community, we will eventually find a cure!!

After each conference, at PSC Partners, we read the post-conference evaluations closely, and try to follow some of the attendee suggestions for the following year. Some successful additions to this year's conference were the well-attended Thursday night casual pizza get-together, Saturday night's karaoke party, our expanded PSC shop and the teen group special activities. We want to address the specific needs of the various groups and are presently working with the quickly expanding pediatric parents group. Keep your ideas coming!

As always, our very generous sponsors absorb half of our conference expenses so that we can offer attendees the conference experience at a reasonable cost. I cannot thank our sponsors enough for their support!

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Scents, Inc.; Yes To; Z Pizza; Media Form; Jeff and
Reggie Belmont, Joanne and Steve Grieme, and the
Pearlman Family

The success of our conference also would not be possible without the support and dedication of all our volunteers. We would like to thank each and every one of you for sharing your time and talents with PSC Partners. The list is too long to mention everyone,

but I'd like to especially thank:

Karen Pearlman, our official photographer, for her [fantastic pictures](#) of the entire weekend

Ken and Mary Ann Long, for heading and organizing all our volunteer teams

Joanne and Steve Hatchett, Joanne and Steve Grieme, Rachel Gomel, Mike Pearlman, Ken and Patty Shepherd, Fred Sabernick, Sandi Pearlman for being our team leaders



Joanne and Steve Grieme and Patty Shepherd for creating and running our classy PSC Shop

Joanne Grieme for putting together our name tags and attendee directory

Rachel Gomel for editing the summaries of all the sessions and all the articles to create this wonderful newsletter edition

Arne Myrabo for taking notes at the General Sessions and organizing and posting all the PowerPoint presentations on our website

Philip Burke, Mike Pearlman, Becky Long, Jeremy Burke, Tiffany Rotondo, Aubrey Goldstein, Ken Shepherd and Sandi Pearlman, all members of our community, for sharing their personal expertise in informative breakout sessions for us

Since we are able to operate as a mainly volunteer organization, we greatly appreciate all of our enthusiastic, hardworking volunteers at PSC Partners who help us by volunteering their time and expertise and/or financially supporting our programs. Congratulations to those who received our 2012 awards:

Partners of the Year:

Dr. Keith Lindor
Dr. Kostas Lazaridis
Dr. Jay Talwalkar
Sandi Pearlman



Volunteers of the Year, given in memory of Pat Bandy
Steve and Joanne Hatchett

Visionary Award, given in memory of Ivor Sweigler
Ken and Patty Shepherd

Appreciation Awards:
Bill Bandy
Ken and Mary Ann Long
The Pearlman Family (Mike,
Eileen, Sandi and Karen)
David and Roz Parry



This year's conference would never have run so smoothly without the help of two very special people in my life: Sandi Pearlman and Meegan Carey. Sandi was my conference co-chair, and it was an absolute delight working out every conference detail with her all year long. Although Sandi is in Rochester while awaiting a liver transplant, she somehow managed to find the energy to get everything done! Nothing would stop her tenacity! Every step of the way, Sandi brought her usual refreshing perspective, and made the planning an enjoyable experience. Sandi is an inspiration to me on how to handle adversity. Even in her worst moments, she does not let go of her sense of humor and continues to give priority to the hundreds of people she touches daily.



Meegan Carey, our Development Director, did an unparalleled job all weekend making sure that everything was working according to plan while doing a lot of trouble shooting in her soothing yet confident manner. Although Meegs is not directly affected by PSC, she is as driven as we all are to offer support to our community and help us find that cure. Meegan is dedicated, bright, proactive, enthusiastic, compassionate, extremely hard working and a wonderful friend. Now that PSC Partners has grown so quickly, we just couldn't exist without Meegan!

Here are a few of my favorite comments from this year's attendees that describe the conference experience:

"The wonderful conference has changed our lives. You will see us at every future conference!"

"What I loved most about the weekend was meeting everyone, the knowledge, the energy, the dedication of the doctors, the friendliness and openness, and the hope."

"I am always inspired by the positive atmosphere and support at the conference. This has become a family and continues to grow!!!"

"Thanks for developing a caring, supportive network for PSCers to turn to when it all seems overwhelming!"

Whether you attended the conference or not, please visit our website where you can access the [PowerPoint](#) presentations from our speakers and also the beautiful conference slide show created by Karen Pearlman.

If you weren't able to join us at the conference this year, please think about joining us at our 2013 conference in Pittsburgh organized in conjunction with the University of Pittsburgh Medical Center the weekend of April 26-28. Join us in learning about the latest developments in PSC treatments and research as well as becoming a part of our PSC Partners supportive family and experiencing the positive energy when we are all together.

I miss that special indescribable feeling that permeates our conference weekend and the incredible friendships that we all share. Thank you to all our participants for making our 2012 conference so memorable!

Here's to a healthy summer for our entire community. Together in the fight, whatever it takes!

Ricky Safer
CEO

Next year in Pittsburgh !

*You're invited to the 9th Annual Patient Conference held
by PSC Partners Seeking a Cure
and organized in association with the
University of Pittsburgh Medical Center
April 26-28, 2013*

Check our website which will be updated periodically

<http://www.pscpartners.org/nextannual>

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We thank our official conference photographers, Karen Pearlman, of K Pearlman Photography and Arne Myrabo for covering the conference with cheer and with great talent. Karen is generously donating to PSC Partners receipts received for photos you purchase. Her website is: <http://kpearlmanphotography.com>. Her conference [slide show](#) and her [conference pictures](#) are on the 2012 conference page!



Foundation Update

by Ricky Safer

PSC Partners Seeking a Cure has four exciting announcements about recent accomplishments:

- As a result of our successful 2012 Road to Research fundraising campaign, we have just met our long-term goal of raising one million dollars that has been restricted for PSC research! A huge thank you to everyone who has contributed to our research fund throughout the years. We are determined to find new treatments and a cure for PSC. Here's to raising our second million dollars for research!
- Starting in 2012, two new co-chairs have been directing the PSC Partners Scientific/Medical Advisory Committee (SMAC). Dr. Richard Green is in charge of our research grants program, and Dr. Christopher Bowlus is heading our patient registry project. Our SMAC has reviewed the 2011 research grant proposals and has awarded \$60,000 seed grants to the three researchers who submitted the most promising PSC research projects. Congratulations to Drs. LaRusso, Baldwin and Weersma. You can read about these studies in Part II of our Conference Edition of our newsletter. You can also find out about [all our research grants](#) to date.
- We are thrilled that PSC Partners has been selected by the Office of Rare Diseases Research (ORDR), a branch of the National Institutes of Health (NIH), to participate in a two year pilot project that will result in a PSC Partners patient registry and that will serve as a model for other rare disease organizations! Our registry will be guided and supported by the leading minds and national organizations that are currently focusing on advancing rare disease research and treatments. For more information about this fantastic opportunity, please read the article in Part II on page 5 of our Conference Edition of our newsletter!
- We are looking forward to our 2013 patient/caregiver conference, which will be held in Pittsburgh in conjunction with the University of Pittsburgh Medical Center the weekend of April 26-28. My conference co-host, Joanne Grieme, and I are already busy planning conference details. It will be a terrific weekend, so please save the date in your calendar and try to join us!

Other important information:

- We would like to welcome two new members on our Board of Directors: Aubrey Goldstein and Roz Parry.
- The DoubleTree Hotel and Suites in Pittsburgh has already set up a personalized link [to reserve rooms](#) for the 2013 conference at our special room rate of \$119 per night plus tax.

- If you would be interested in being a co-host for the 2014 conference, please [fill out the form](#) and return it to contactus@pscpartners.org before our January 15, 2013 deadline.
- If you are a researcher who would like to send in a grant proposal, please go to www.pscpartners.org/apply and submit your proposal before Monday, September 3, 2012.
- Please think about holding an individual fundraiser in your area as part of our Save the Day campaign. Save the Day is up and running and events are already in progress. Join in or create one of your own. It's easy, fun and we're here to help you get approved and get going on the quest for a cure! Want to hold your fundraiser the same day as PSCers all over the globe? October 12-14 is the time for our collective effort to educate our local communities about PSC and to raise funds to support the mission of PSC Partners Seeking a Cure! For more information or to see what's happening, turn to www.pscpartners.org/fundraisers#Save-the-day for updates and more.
- If you have any ideas for our foundation or would like to volunteer, please write to us at contactus@pscpartners.org.

FROM DR. JAY TALWALKAR ON ATTENDING THE 2012 CONFERENCE

As a first-time attendee and speaker at the recent PSC Partners conference in Rochester, MN, my experience was absolutely outstanding. The work done by this organization and its tireless leader Ricky Safer is absolutely essential for our continued efforts in finding a cure for PSC.

The most remarkable experiences I had during the conference were the one-on-one interactions with the attendees. I was absolutely impressed and humbled at the level of knowledge and understanding they displayed. Furthermore, I was thoroughly engaged and excited by the insightful questions each person had about PSC.

On behalf of our group here at Mayo including Drs. Lindor and Lazaridis, I am proud to say that this weekend is one we would like to share again with all of you in the near future !

Jay Talwalkar, MD, MPH, Hepatologist, Mayo Clinic Rochester

The Love Letter

by Sandi Pearlman

Here's the thing, I don't know anybody who doesn't truly, secretly, proudly and completely adore love letters. I mean, what's not to love? A love letter is emotion in its truest form. It's thought combined with action and the care it takes to put pen to paper or fingers to keyboard. It's somebody taking a minute (or 10 or 20) of their time to profess their innermost appreciation for another. There isn't a person alive who can resist the allure of love no matter how hard they may try. And for me, for many, PSC Partners conferences are a love letter of sorts. It's all of us, taking time from our overly busy and hectic lives and putting down obstacles and illnesses and demands to be with each other come what may. For we know, this all-too-short weekend is our chance to profess our love and dedication to one another and our commitment to curing not just ourselves, but all of us. We're together in the fight, whatever it takes, not because it's the PSC Partners slogan; but because it describes what's written in our hearts and souls, in our very cells. It is the essence of a love letter, eternal, beautiful, powerful.



I've been accused of overstating the amount of love in our group. I've been told that what I'm saying simply can't be true. But you know what I hear most on the subject from first-time attendees and those doubters, I hear that I was right. I hear their joy as they tell me they now know I wasn't overstating, selling them flowery promises with wilted stems at the core. There's simply no such thing as isolation at the conference. From the hugs and greetings as one enters the door to the handholding and tissue sharing in breakout groups or as we listen to sometimes dire statistics, the love is there. It bubbles out in song--lots of songs this year--and in stories and in shared moments. It's there in the knowledge that our loads are much lighter because we have everyone's shoulders to share the weight and it's no longer our battle alone.

As anyone who has ever been in love can attest, love doesn't just come straight at you charging like a bull in a china shop. It caresses, like words on a page, until you're so enveloped you can barely remember what life was like before. And once you're open to the love and beauty in the world, it pours forth, even from unexpected sources. At this year's conference alone, there were doctors who were mobbed like rock stars as they patiently answered questions and listened to stories. We loved them and their time, expertise and attention and they proved again and again that they're more than physicians. They showed us in words and in actions that they're comrades in the fight, aiming to cure PSC and stop its ravaging effects before it's too late. Then there are our sponsors, some who attended and others who couldn't, who give of themselves to support us both in spirit and in funding to allow us to keep our conferences accessible to so many. And there are all of us, PSCers and caregivers from all corners of the world. We come together to make ourselves more informed, more connected and to share of ourselves and our experiences with those who understand us most, those who are just like us; those who know the initials PSC like we know the back of our hands and who know that we're more than a diagnosis, even when our bodies try to prove the very opposite.

This was my fifth conference and, for me, the love just gets stronger and stronger. It defies language barriers and sometimes grim information. It doesn't matter if we've known each other for five years or five minutes. As Renée Zellweger famously said, "You had me at hello." PSCers, caregivers, you all had me at hello. You make me feel inspired and brave. You remind me that I not only can survive PSC, but I can thrive with PSC; that we all can.

The thing about love letters is that they stay forever etched upon our hearts and minds long past the moments we first encounter them. The words and feelings imprint themselves in our memories and in our cells becoming a part of us that we can call upon when things get rough. Our conferences may only be one all-too-short weekend a year, but as anyone who has ever attended knows, they are a love letter that keeps on giving, bringing us back to special moments and the love shared time and again. They're love letters that we keep tucked away in our hearts until we meet again, written in the most beautiful script and forever meaningful. Sending love until we meet again in Pittsburgh.

Forever yours,
Sandi



The Annual Conference Experience

*We asked seven participants at the conference to share their experiences.
Most were first-time attendees.*

The Family I Never Knew I Had

By Martin James Frier

Ever since I was 9 years old, I have had to deal with PSC, this terrible chronic illness. I remember my doctors telling me it was a rare liver disease. I thought to myself, "Great, I'm the only person who's going through this, and there is nobody I will ever be able to relate to."

Up until I was 18, I never knew of or met anyone who had gone through what I have had to go through - at least not until I discovered PSC Partners. PSC Partners has truly been a blessing to me. Ever since I attended the 2012 conference in Rochester, I started to have hope that there will be a cure for PSC. I have met the greatest, most positive people I have ever met in my life. I was

shocked: Here are a bunch of chronically ill patients who are more positive than most healthy people. It was great being able to tell my stories, to listen to other stories, and to be able to relate to their trials and tribulations and say, "Oh, yeah, I feel the same way," or "Yeah, I can't stand itching all the time." It was like a family I never knew I had until now. I will definitely attend every conference I possibly can.

Which is Why I Will Be Back Again Next Year

By Stephen Harris

As someone who was diagnosed with PSC during the first month of medical school, my day-to-day environment soon provided me a first-class education in the physiological implications of having the disease. But it was only after I attended the 2010 conference in Hartford CT that I realized there was more to PSC than LFTs and CBCs. By the end of that weekend, new friends had unscrewed my head for a while and encouraged my heart to have a say in things.

Now, two years (and a small condo's worth of Remicade) later, I'm in the lobby of a Rochester hotel, surrounded by familiar faces. The conversation is comfortingly open. "How is Sandi doing?" "Does anyone know what time Dr. Talwalkar is speaking?" "I just met this girl. How do I tell her my liver looks like a *Grimanesa Amoros* bubble sculpture?" (Don't ask!)

As always, this year's conference was a fantastic mixture of humanity and medicine, homecomings and high-intellect, and above all, self-empowerment and self-education. Many things we don't have an equation for hold equal court with science here. Whether your concern is the molecular biology of bile duct epithelium, or how to persuade your loved one to take their medication on time, there is undoubtedly someone in the room who can relate, and help you sort it out. Me? I just wandered about aimlessly, missing breakout groups, eating free food, and being reminded that there is more to this PSC community thing than I think.

Which is why I will be back again next year.

Pittsburgh Here I Come!

Marleen from Holland

Four weeks after my PSC diagnosis, I was packing my bags and getting on a plane to attend the PSC conference I had found out about at home in Holland. Little did I know that this event would be a life-changing experience for me, that I would be returning home with tools and insight to deal with PSC in the best possible way.

It wasn't only information I received. I also got good pointers on how to deal with day-to-day issues brought on by PSC. Some of the information I received, and which I then thought I didn't need to hear, has turned out to be extremely helpful to me.

It was amazing that the conference was organized by volunteers only and was so professional and in such a great location. I was amazed to see the interesting variety of subjects and the top-notch quality of the speakers. Yet, it was remarkable that the personal touch was tangible in every step of the way. Along with the occasional tears, there was also tons of laughter.

I travelled over 4,000 miles to get to the conference. I never felt lonely, not for a second. The event was a brief three days, but I ended up with friends for life.

Meeting people who are linked to PSC in one way or another was of great value to me. The conference gave me a huge support system comprised of people of all kinds who instantly understand me and who are always there for me 24/7. Yes, I can safely say that I no longer feel alone with my PSC.

The conference provided me insight not only into the patient's perspective but also into that of the caregiver. I found this angle extremely helpful because I can now cooperate and interact much better with my caregivers.

I would wholeheartedly advise anyone closely connected to PSC – not only PSCers, but also caregivers, family members, doctors and others in the medical field – to attend the annual PSC conference.

I have already marked next year's conference date, April 26-28, 2013, on my agenda.

Pittsburgh, here I come!

From a True Friend of PSC Partners

by Eileen Solomon

(Eileen is Director of Special Events at the Mount Sinai Medical Center in NYC. She and Stephen Harris run the Mount Sinai PSC support group. At our conference they interviewed several of our speakers and attendees. You can hear their [interview with Dr. J. Talwalkar](#) recorded at our conference.)

This was my second PSC conference. The feeling of warmth, support, grace, and a willingness to be open, vulnerable and strong permeated the entire weekend. The conference was filled with love. The energy that the entire team put into making this annual gathering happen was extraordinary and felt by all.

Details: Nothing was too small. No one was left out, alone, or unattended. Timing: precise and mindful of the speaker and the audience. Nourishment: abundant, and I don't just mean the food, which was really good. The amount of information exchanged between individuals and the professionals was fabulous. I continue to learn about PSCers and am inspired by their strength. Thank you for letting me be a part of this awesome group.

Warmly,

Eileen

Planting Friendships

By Chelsea Yaeger

It was a wise man who once said, "... friendship is precious, not only in the shade, but in the sunshine of life, and thanks to a benevolent arrangement, the greater part of life is sunshine."* I like to think of friendship as a plant, or many different plants, rather. Most dry out. Some turn out to be weeds. Some grow and then suddenly wilt and fade away, back to the mulch from which they were created. Only a few flower and remain.

I am blessed to have had so many new friendships grow from little seeds at this past conference. Some even skipped the sapling stage, such as my friendships with Katie and Aly.



My friendship with both girls shot up immediately and, within a few hours, we were giggling and had so many inside jokes already. I honestly don't think we were apart for more than an hour (besides sleeping time). I'm really surprised we didn't have sleepovers in each other's hotel rooms!!! Between singing Spice Girls on Karaoke Night, the little field trip we took, the workout room with our friend Swole (you know who you are), late nights in the comfort room, pressing all the buttons in the elevators, and eating lots of sugary cookies (no soda this year, guys!!), the three of us connected instantly.

I do think having PSC bonded me with both girls. Katie, because she went through almost exactly what I did with PSC, but she's better now. Last year when I met her, we were going through exactly the same thing. Aly, because I was her mentor (yay!) and because she, too, knows what it's like to feel different, to take medicine, and to have to consider PSC and its effects in college choices as well as life choices. I feel as though we were thrown together by this awful disease, but am I glad we were!! Things have a funny little way of working themselves out.

Aly and Katie are those rare kinds of people to me – those friends that last even though you don't always see them, because you know they're always there. As Pam Brown once said, "A friendship can weather most things and thrive in thin soil; but it needs a little mulch of letters and phone calls and small, silly presents every so often - just to save it from drying out completely."

*quote by Thomas Jefferson

Even If You Fall Asleep

By Matthew Buddha Ayuen

After attending the 2012 PSC Partners, I have nothing but great things to say. It was my first year attending the conference. As my parents were there in 2011, they assured me that I would have a great time. Despite being skeptical, I figured that at the very least I would learn a lot even if I didn't enjoy myself. In the end, my parents could not have been more right!

It was such an unbelievable experience, meeting so many people whose lives have also been affected by PSC. Everyone, from PSCers to their caregivers, siblings, even PSCers' children participate in this event, and seeing how different everyone's stories are makes the experience somehow surreal.



Why was the conference so great? Well, the conference is a place where even if you fall asleep during one of the speaker presentations, which I did ... oops, people understand. It's a place where no one asks you, "Why are your eyes yellow?" or "Why do you itch so much?" Furthermore, you learn so much throughout the conference, and the information comes from everyone. For instance, I heard how different people dealt with certain aspects and

symptoms of PSC. I also learned a great deal of technical information from Mayo doctors - how to interpret blood tests and about medication.

Despite all that I learned about PSC, the best part of the conference for me by far was meeting all the amazing people. Everyone was so fun loving and nice that it almost seemed too good to be true. On top of being super informative, I found the conference really therapeutic. As an eighteen-year-old, it was great to share and discuss with other young adults of my age what my last ERCP or liver biopsy was like.

If I can leave you with a thought, it's this one: I left the conference feeling many emotions. Above all, I left happy. The whole weekend made me feel as if I had left behind all my worries even though I was facing them straight in the face. I was also sad, sad that it was over so quickly. I guess time flies when every minute is meaningful and fun. Finally, I was excited. I was excited to go home and tell my doctors everything I'd learned. Most of all, I was, and still am, excited for the next PSC Partners conference in Pittsburgh. It will be great to see everyone again, and I really hope to see you there, too!

I Found My People

By Ruth Beinart (South Africa)

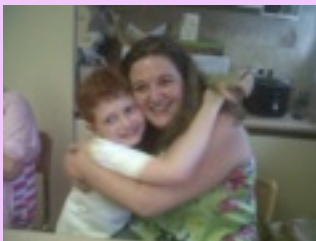
So ... when sitting down to write about the conference, I thought to myself, if I could think of a single word to describe the 2012 PSC conference, what would it be? It took about three seconds – FAMILY.

In one weekend, I met the family I never knew I had. I walked into a room of people who all spoke the same language as me (PSC). Everyone had the same worries, same heartache, same frustration, fear, anxiety, hope, anger, determination and unwavering mission.

When Dylan was first diagnosed in 2007, my whole world fell apart. I was scared and alone. His doctor was a very knowledgeable woman but was unapproachable. I tried to find out as much as I could and be as informed as possible. The information available on the Internet was insufficient and overwhelming.

I started to ask questions. Why was he on this medication, what did this blood test mean, why is he so tired, is this itching normal, can he take pain killers if he has a headache, what about medication for the common cold? I had so many questions and no one could really answer me.

Our pediatrician said that he could no longer help us; even a cold took on major dimensions; what started as a minor break in his arm ended up in a cholangitis attack, and no one was sure about anything. No one had any answers.



I was frustrated and angry. I wanted more. More answers, more advice, more people who understood what this disease was about, more information, more options, more hope. That was when I decided to attend the conference. I needed help.

I still remember when I first walked into the room at the conference. I was enveloped in welcome. Everyone was so excited to meet me and so pleased I was there. I have never felt so comfortable so fast amongst a group of strangers.

I had found my people. I could ask anyone questions and get such great advice and support. I was able to share my story without sounding dramatic. I met other moms who knew what I was feeling.

Every aspect of the conference was fantastic, from the wonderful coffee, to the incredible speakers that gave of their time and knowledge. I found out about medications I had never heard of before. I got tips on how to give liquid medication. I found out about some supplements and vitamins. I learnt how to read my son's blood tests and what to look out for. I got information on the variety of different scans and procedures for checking PSC and the ones that were best to use in different situations. I made contact with a variety of specialists and professionals who gave me great advice on international medical care and on how to proceed. I finally feel I am steering this ship rather than being a passenger on a very scary drive.

I walked away from the conference with a course of action in my head. I feel I have some sort of direction to go in and more confidence to insist on what I want for Dylan and what I think he needs. I have more knowledge about what direction to take and what care is available. I can already see that I am able to make better-informed decisions.

Thank you to everyone, firstly for the amount of work that went into the conference, secondly, for the care and respect that was given to everyone, and thirdly for your understanding and for giving of yourselves.

The weekend was amazing and overwhelming. I felt joy, love, confusion and fear, panic and confidence, hope, strength and power, courage and knowledge. But most of all? ... I felt FAMILY.

YOUR EXPERTISE NEEDED!

Do you have online newsletter publishing experience? We need your expert advice!

rachel@pscpartners.org

All the speakers were true experts on PSC. Our Mayo doctors are all world renowned experts. Philip and Aubrey....What more can I say? I truly felt that everyone was in our corner.

It was really nice to have someone contact us to be a mentor before the conference, but this group is so friendly that you feel everyone is your mentor!

Conference Session Summaries

Reported by Arne Myrabo

Management of Symptoms Related to PSC

Jayant A. Talwalkar, MD, MPH, Associate Professor of Medicine
Mayo Clinic Rochester

Presentation slides available at [Management of Symptoms Related to PSC](#)

Reported by Arne Myrabo

Overview. PSC is a chronic cholestatic (obstructed bile flow) liver disease of unknown and probably varying causes. Often associated with inflammatory bowel disease, it is characterized by diffuse, chronic inflammation which frequently results in fibrosis (scarring) of the bile duct tree. This can involve all areas of the liver and/or the extrahepatic (outside the liver) bile ducts. The scarring within the liver results in cirrhosis, which can progress to portal hypertension (variceal bleeding), ascites (fluid retention) and jaundice (yellowing skin). **Inflammatory Bowel Disease.** In the past, ulcerative colitis had been associated with PSC in about 80% of PSC patients. Due to earlier detection, current estimates are

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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.

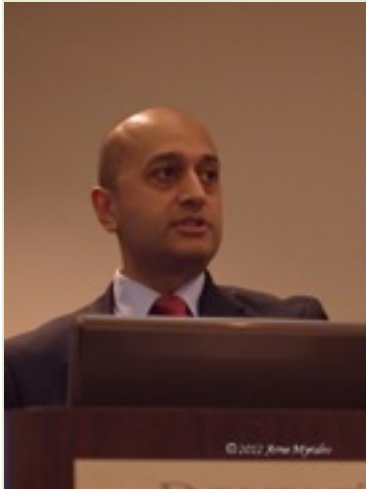
60-80%, that is, more variable than previously believed). IBD is also seen in association with Crohn's disease, but usually only in those cases with colonic involvement (PSC with small bowel involvement only is uncommon). The prevalence of UC is estimated to be about one in five hundred people.

Symptoms. At diagnosis, 15-45% of PSC patients are asymptomatic. A history of jaundice, abdominal pain and fatigue are seldom reported at initial diagnosis now. For those that are symptomatic at diagnosis, fatigue and pruritus (itching) are the main complaints.

Liver function tests (LFT). Alkaline phosphatase (ALP) is nearly always elevated. AST/ALT are considered to be inflammation markers and are often elevated. Bilirubin, albumin and prothrombin time are usually normal at

diagnosis. Some patients have fluctuating levels with no symptoms, but a cholangiogram (bile duct tree image) almost always reveals features characteristic of PSC. Persistently normal LFTs do not mean that there is no PSC.

Cholangiography. In the past, Endoscopic Retrograde Cholangiography (ERCP) was used to diagnose PSC. Advances in the non-invasive imaging technique magnetic resonance cholangiography (MRCP) have currently relegated ERCP to an



interventional technique. Stronger magnets and 3-D imaging can create extremely detailed images of the biliary tree. Percutaneous cholangiography is rarely used for imaging.

Liver biopsy. Liver biopsy may not be required in all cases, but it is needed to exclude autoimmune hepatitis (AIH) overlap, and to confirm small duct PSC. Small duct PSC is a variant of

PSC, but it is managed in the same way as PSC. A biopsy that shows concentric fibrosis around bile ducts is confirmation of PSC.

Fatigue. Fatigue is one of the more common symptoms of PSC, and it occurs in up to 50% of patients. There is no known correlation with age, sex, biochemical tests (LFTs) or [Mayo Risk Score](#). Mild disease may exhibit debilitating fatigue, and advanced disease may have no fatigue. No effective therapy has been identified, but there is some indication that selective serotonin re-uptake inhibitors (SSRIs) may be of benefit. Modafinil, which is used for narcolepsy, also may have some benefit, but side-effects and restrictions on use make this medication problematic.

Pruritus (itching). The frequency of itching is between 20-60% of PSC cases. If there is a dominant stricture, balloon dilatation may dramatically improve symptoms. As with fatigue, there is no correlation to

disease stage, and the cause for this symptom remains unknown. The treatment algorithm at Mayo is Cholestyramine, Urso, Sertraline (SSRI - Zoloft), Rifampin and occasionally, Naltrexone. Urso has had variable success, usually only with mild itching. Sertraline is well tolerated and does seem to work for some. Rifampin may show dramatic improvement, but there are possible side effects that must be monitored.

Metabolic Bone Disease. Osteoporosis is much more common than vitamin deficiency. Hormone replacement therapy may be suggested for women. Calcium with Vitamin D is helpful, and bisphosphonates may be helpful. For those requiring steroid therapy, bone disease may worsen and must therefore be monitored carefully. Calcitonin has not been shown to be helpful.

Biliary strictures. Typically managed with ERCP (seldom with percutaneous cholangiography), dilatation alone is usually adequate for dominant strictures. In the few cases where stenting is required, it is preferable to leave in the stents for no longer than 4-6 weeks. It is not unusual for a stent to need to be placed only once. Antibiotics are given post-procedure to limit possible infections.

Medical Therapy. Numerous therapies have been tested, but very few have shown any promise. There is still great interest in Ursodiol, although high-dose (28-30 mg/kg/day) is no longer recommended. Since those with UC and PSC have an increased risk of colorectal cancers, an annual colonoscopy is recommended.

Cholangiocarcinoma. Cell brushings should be taken during an ERCP to test for abnormal cells. Cytology alone has a sensitivity of 4-20%, but fluorescence in situ hybridization (FISH) is more sensitive (35-60%).

Management of PSC:

Clinical visit every 6-12 months

Serum liver tests every 3-6 months

Ultrasound / MRCP and CA 19-9 test every year

Colonoscopy every year

Bone density test at diagnosis and every other year thereafter

Vitamin levels – only if total bilirubin is elevated or there is cirrhosis

We thank **Arne Myrabo** for setting up our 2012 conference website. You can access the presentation slides and other information for the 2012 conference at <http://www.pscpartners.org/PSCConf12/index.htm>



Coping with PSC: It's Not "All in Your Head"

Philip Burke, PhD, Clinical Psychologist, Assistant Professor of Clinical Psychology,
Southern Illinois University, Carbondale, PSC Patient

Presentation slides available at [Coping with PSC: It's Not "All in Your Head"](#)

Reported by Arne Myrabo



Arne Myrabo noted that his summary did not do justice to the compassion and humor that permeated Dr. Burke's presentation.

Understand the Need to Cope: Physiological Basis for Need to Cope with Chronic Stress

We tend to focus on the biological components of PSC for several reasons. Many of the typical PSC symptoms are not specific to PSC. For example, fatigue and itching are the two major patient complaints with PSC, and yet there are many conditions that can result in those vague symptoms. The symptoms can also be very erratic. There are people who have PSC for over 30 years and are

still doing well. There are also those who receive a transplant 1-2 years after diagnosis. There are some who have mid-20s MELD scores but still feel fine. There are those that have fairly low MELD scores and yet are on disability. It is much easier to explain a cholangitis infection which has a clear, biological reason behind it.

We do have a biological need to cope. Yes, it's NOT all in your head; there IS a physiological component. We are built to respond to physical and psychological demands (flight or fight response). When temporary, our acute stress response system helps us meet demands. In order to facilitate rapid decisions in preparation for threats, *acute stress*

- narrows our thinking
- focuses attention
- restricts emotions and
- limits creativity

But when this response is extended, as in a chronic illness, the response takes its toll. The acute stress response model does not fit PSC since the threat is unpredictable and the action one must take is uncertain. The result is chronic and recurrent stress. A different coping strategy is needed in dealing with chronic stress.

In the case of PSC, "Don't just do something, sit there," might be the better response.

Shift to a Bio-psycho-social-behavioral Model of Health

In reality, the components of biology, behavior, psychology, sociology and health are interrelated. You cannot address them individually. And there is much more to PSC than taking medications.

An example is mild to moderate itching or pruritus (note that this does not apply to intense itching). If you can apply good coping methods at the time itching is first noticed, you may be able to break the cycle and at least endure/tolerate the itching. Calming strategies may help to reduce the arousal of the fight/flight response, allowing you to concentrate on something else (or be distracted), reducing the focus on itching. For mild to moderate itching, medications are a big part of this process – but they are not the *only* part.

Some examples of coping strategies:

Balanced diet

Regular exercise

Regular sleep schedule

Medications/supplements

Deep relaxation

Financial/insurance planning

Balance: rest with “Pushing On”

Re-Prioritizing/Benefit-Finding

Rethinking demands as priorities

Problem-solving (for solvable problems)

Practicing acceptance

Seeking social support

Recognizing mental health problems as expected parts of illness & seeking help

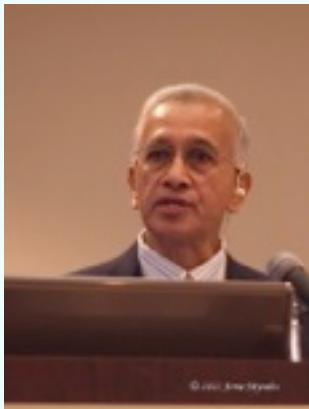
Reducing self-blame while accepting responsibility for own health & healthcare

Being a “team leader” for healthcare

Reducing blaming others

Visit <http://www.morethanillness.org/> for copies of information presented.





MELD System: Past and Present

Patrick S. Kamath, MD, Professor of Medicine,
Mayo Clinic College of Medicine

Presentation available at [MELD System: Past and Present](#)

Please note that Dr. Kamath's slides are quite comprehensive.

Reported by Arne Myrabo

What is the MELD Score?

MELD stands for Model for Endstage Liver Disease. It is a mathematical representation of the risk of mortality at three months. It is entirely based on three routinely obtained blood tests: INR for prothrombin time, serum creatinine and serum total bilirubin. The calculation weighs INR more strongly than creatinine, and creatinine more strongly than bilirubin. Examples of three-month mortality risk are 10% at a MELD of 22, 30% at 29, 50% at 33, and 80% at 38. An online calculator is available at <http://www.mayoclinic.org/gi-rst/mayomodel6.html>.

Why was liver allocation changed to the MELD-based system in the US?

There are two philosophies on allocating organs; prioritize those who are sickest, or those who benefit the most. In the US, the sickest patients are transplanted first. The question rests on how to determine who the sickest patient is.

The previous allocation scheme favored those who were on the waiting list the longest, and those in ICU. Length of time on the waiting list became the main determinant, and the system could be "worked" by ICU placement. In 2000, the Department of Health and Human Services issued the Final Rule Mandate on organ allocation. It stated that:

- Priority for organ allocation should be established on objective, measurable, and clinical criteria
- Waiting time must be de-emphasized
- Patients should be rank ordered on the liver list according to predicted mortality

The major changes were that the medical status (1, 2A/B, etc.) was replaced by probability of death, waiting time was no longer a consideration, and no preference was given to ICU patients.

How is MELD used for organ allocation?

- Fulminant hepatic failure has the highest priority
- The highest MELD score determines priority amongst patients with cirrhosis and same blood type
- Waiting time is used only to break ties at identical MELD scores
- MELD scores are updated at regular intervals

How has MELD affected liver transplantation?

The number of recipients in ICU at transplant is about half of what it was before MELD. The number of new listings has decreased; the number of cadaveric transplants has increased; and the number of

deaths on the waiting list has decreased steadily. The median time to transplant has decreased from three years to about one year. Unfortunately, the post-transplant survival has not improved, and remains the same.

There is some risk in transplanting at too low a MELD score. In patients with MELD below 14, there is a higher risk of 3-month mortality with transplantation as compared to 3-month mortality without transplantation. In patients with MELD between 15-17, there is no survival advantage or disadvantage with liver transplantation. Survival advantage is seen only once MELD scores are higher than 17 with the greatest advantage being seen at higher MELD scores.

Overall, MELD-based liver transplantation shows advantage for those with high MELD score, renal failure, anticoagulation, hepatocellular carcinoma, or for some special diseases and conditions. **It disadvantages those with low MELD scores but debilitating symptoms such as extreme fatigue and pruritus, ascites, cholangiocarcinoma, encephalopathy and recurrent cholangitis.**

Summary

The MELD system is an excellent predictor of pre-transplant survival, and has resulted in:

- Decreased registrations
- Decreased death rate on waiting list
- Sicker patients being transplanted
- Unchanged post transplant survival
- Better defining survival benefit and optimal timing
- Evidence-based decision-making

A Question and Answer period followed Drs. Talwalkar, Burke and Kamath's presentations.

The presentation slides can be accessed at <http://www.pscpartners.org/PSCConf12/index.htm> and the audio recordings of the Q & A period, at <http://pscpartners.org/PSCConf12/sessionIqanda.htm> (session I) and <http://pscpartners.org/PSCConf12/sessionIIqanda.htm> (sessionII)

NOTE TO RESEARCHERS:

The deadline for the PSC Partners Grant Program applications is September 3, 2012.

<http://pscpartners.org/apply>



Ongoing and Future Medical Therapies for PSC

Keith D. Lindor, MD, Executive Vice Provost for Health Solutions, Arizona State University, past Dean of the Mayo Medical School

Presentation slides available at [Ongoing and Future Medical Therapies for PSC](#)

Reported by Arne Myrabo

High Dose Urso Study

Urso had been shown to be useful in PBC (primary biliary cirrhosis), and a small trial with PSC showed promise, especially at higher doses. Due to unexpected adverse effects (primarily an increase in liver transplants and varices), the study was terminated. This clearly shows the need to perform these large-scale studies and serves as a cautionary tale for new treatments that seem promising.

One clear benefit is that Urso reduces the risk of colon dysplasia (at high doses), but two of three previous studies at “normal” doses only suggest a small benefit.

A recent study shows that normalization of ALP (alkaline phosphatase) levels improves outcome in PSC. It is interesting, and perplexing in that it doesn't matter how it gets normalized – whether spontaneously or by using Urso. It is possible that there is a subgroup of PSC patients that *does* respond to Urso and benefits from this medication.

In summary, we really don't know what to do with Urso at this time. 25-30 mg/kg/day is clearly too high, and the 13-15 mg/kg/day used in PBC doesn't show an overall benefit in a study done 15 years ago. To add to the confusion, the PSC practice guidelines from the American Association for Liver Disease (AASLD) and European Association for Studies of the Liver (EASL) differ. AASLD says no URSO under any circumstances. EASL doesn't recommend low or high dose and states that there is insufficient information for moderate dose.

PSC - A, B, C...?

There may be different variants of PSC which may require different treatments. Fifty years ago we had hepatitis. Now we have six different hepatitis diseases, each of which has different treatments. There may be an analogy with PSC.

For example, 10 percent of PSCers have elevated IgG4 serum levels. The risk of transplantation for PSCers with elevated IgG4 is much greater, and the need for transplant arrives sooner than the

average. Unlike most PSC, this type of PSC *does* respond to steroid therapy. Budesonide (steroid) has not been shown to be useful in treating PSC, but if the IgG4 data in the study is reviewed, it can be observed that two out of three with elevated IgG4 had dramatic responses.

We need to understand subtypes to identify treatments.

Bacterial theory

Adult Vancomycin study results are currently in preparation. The results are not as impressive as the pediatric study was. Again, it appears that there is a subtype that does fit into the anti-bacterial theory of PSC. A Finnish Urso study showed slight improvement with the addition of Metronidazole. Other antibiotics being investigated are Tetracycline and Azithromycin. Once we learn more about the microbiome of PSC, we may be able to tailor treatments accordingly.

Immunologic Therapies

We also need to understand the genetic side of PSC, especially as it relates to the immunologic aspects. Broad spectrum immunosuppression has had limited success. Tacrolimus showed dramatic improvement in one study, but the results could not be replicated. Additionally, side effects were substantial. As stated earlier, the IgG4 variant of PSC responds to steroid therapy.

Other Promising Therapies Under Study

- Fenofibrate – Dramatic improvement was observed in ALP. Currently, funding is being sought for a large-scale randomized study.
- Obeticholic Acid (INT-747) – There were improvements in ALP with side effect of itching at high doses.
- Xifaxin (another antibiotic, also used to treat hepatic encephalopathy)
- norUrso – This medication is just going into clinical testing with PSC patients.
- All-trans retinoic acid study is currently in progress.
- Milk thistle – This product is showing promise and is being revisited.
- DHA (fish oil) is also showing promise.
- Antifibrotics (reverses fibrosis) – Numerous proposals are in the works.



Dr. Lindor receiving
Partner of the Year Award

BREAKING NEWS!

NORD announces the FDA Safety Innovation Act which has wide implications on PSC and all rare diseases.

<http://www.rarediseases.org/news-events/news/fda-safety-innovation>

PSC Partners is a member of NORD



NORD
National Organization for Rare Disorders
2011 National Organization Member



PROGRESS: Beginning to Understand the Genetic Predisposition to PSC

Konstantinos N. Lazaridis, MD, Associate Professor of Medicine, Associate Chair Research, Division of Gastroenterology and Hepatology, Associate Director, Rochester Center for Individualized Medicine
Mayo Clinic College of Medicine

Presentation slides available at [PROGRESS: Beginning to Understand the Genetic Predisposition to PSC](#)

Reported by Arne Myrabo

PSC remains a problem that must be solved. Many pieces are still missing. In 1924, Delbet described the first case of PSC. The first PSC case series reports were published in the early 1980's (R. Wiesner, N. LaRusso of the Mayo Clinic and R. Chapman in the UK). The cause of PSC remains unknown and there is no medical therapy available.

PSC is probably caused by a combination of a genetic predisposition and an unidentified environmental trigger. There is a recognized overlap with IBD. It is estimated that there are about one million cases of IBD in the US, and 30,000 cases of PSC. Seventy-five percent of those with PSC have IBD. Ultimately, we need to figure out how to identify the IBD patients that will develop PSC.

PSC is a heterogeneous disease with more than one variant. As a result, different genes, and probably different environmental triggers are involved. If the genes and triggers can be identified, new diagnostic capabilities, disease progression assessments, therapies and even preventive measures may be developed.

PSC Resource of Genetic Risk Environment & Synergy Studies (PROGRESS) was developed in 2005 to better understand the cause(s) and pathogenesis of PSC, in addition to improving PSC prediction and therapy. (*editor's note: A PSC Partners seed grant garnered this study a \$3.5 million NIDDK/NIH grant.*)

The PROGRESS study involves completing a questionnaire and giving a blood sample. Liver transplant recipients are not excluded, and there is no need to visit Mayo Clinic to participate. For some participants, selected relatives may also be recruited. Currently, the study has about 1000 participants from nearly every state. Recruitment at various medical centers has increased participation.

The specific goals of the NIDDK grant are to expand the number of participants, complete Genome Wide Association Studies (GWAS) for PSC & UC, including replicating results, and determine environmental risk factors using questionnaire data.

Current research examples include MMP3 research, immunochip experiment (international – 4500 participants) and family exome sequencing.

There is serious interest in finding families with multiple members affected by PSC, IBD and gallstone disease. Questionnaires and genome analysis were used to identify specific genes involved in this very rare variant of PSC (R595X mutation in ABCB4). Exome or Whole Genome Sequencing in the near future, in addition to identification of environmental triggers, will improve the diagnosis and therapy of PSC.



Sleep and Chronic Illness

John G. Park, MD, FCCP, FAASM
Assistant Professor of Medicine
Division of Pulmonary Critical Care and Sleep
Mayo Clinic College of Medicine

Presentation slides available at [Sleep and Chronic Illness](#)

Reported by Arne Myrabo

There are very few studies *specific* to sleep and PSC. However, many of the techniques used to address sleep issues apply to PSC as well.

Normal sleep

Normal sleep is comprised of numerous periods of wakefulness, minor sleep, rapid eye movement (REM) and deep sleep. Deep sleep changes over the course of the night. We usually see more deep sleep in the first half of the night. REM sleep is shorter during the earlier part, but longer in the latter part of the night. The “normal” pattern also changes as we age. Wake After Sleep Onset (WASO) normally increases with age, and deep sleep duration decreases as we age. On the average, probably we need about 7 – 7.5 hours of sleep per night. Most people get about 6.5 hours of sleep during the week. It is normal to have about 20 – 30 minutes of wake time after falling asleep, and to see up to 15 – 20 arousals/hour of sleep. This should give you an idea of whether your sleep patterns are normal.

Chronic diseases and sleep

Sleep Deprivation – This term is defined as insufficient sleep duration to support a full level of functioning the next day. In addition, there may also be poor *quality* of sleep. Most people in the US are sleep deprived, and try to make it up on the weekend. It may take more time than the weekend to catch up to your sleep deprivation.

Fatigue – It is not the same as sleepiness. With fatigue, one may feel weak, and have difficulty concentrating and initiating or maintaining activity.

Sleep and IBD – 50 percent of ulcerative colitis and Crohn’s disease patients report substantial fatigue and 20-25 percent report chronic fatigue. This can be caused by disease symptoms, lowered hemoglobin (anemia) and/or altered sleep. Sleep interruptions may be due to disease severity (flares), urination, bowel movement, breathing, hot/cold, bad dreams, anxiety or perceived stress.

Predictors of poor sleep – First comes poor health, second depression, then alcohol use.

Sleep and Depression – 6-8.5 hours of sleep is optimum. Less or more sleep can increase symptoms of depression.

Fatigue and PSC - The amount of fatigue does not correlate with the severity of PSC, but is thought to be related to GI symptoms and depression. Cirrhosis delays the circadian rhythm by 4-6 hours.

Disease severity, hemoglobin (anemia), and altered sleep are the main contributors to fatigue. The disease itself often causes interrupted sleep. Co-existing depression will also affect sleep negatively. A 4-6 hour *delay* in circadian rhythm is observed in patients with cirrhosis.

Impact of poor sleep

Disrupted sleep affects deep sleep and REM sleep the most. Sleep deprivation results in change of mood, decreased quality of thinking, accidents, inferior quality of life, weakened immune function, decreased appetite and cardiovascular function. Continually interrupted sleep can have the same effect as going without sleep for 50-60 hours. Interrupted sleep also *decreases* pain tolerance. That is why more pain can be experienced with poor sleep.

Treatment

Treat the underlying disease and co-existing sleep disorders (restless leg syndrome, sleep apnea, etc.)

Practice good sleep hygiene – As you would with children, slow down, dim the lights, and avoid stimulating activities. Plan for adequate sleep duration. Minimize caffeine and alcohol, television, and reading. Beds are meant for sleeping. If you see that you can't get back to sleep, step out and do something quiet. Read something short (and boring), and don't watch the clock. Short naps are okay during the day, while long ones may affect your "normal" sleep time.

Exercise – 30-60 minutes of moderate exercise several times a week improves sleep quality and duration. Start slowly and use moderate intensity, even with strength exercise.

Cognitive behavior therapy – This tool may improve sleep quality and duration.

Medications – Those that have shown improvement in sleep quality are Pregabalin and Trazodone (from fibromyalgia management), Duloxetine (from depression management), Hydroxyzine (histamine blocker), Sodium Oxybate (narcolepsy).

Alternative medicine – Studies show that acupuncture/acupressure may improve insomnia. In addition, yoga and spa therapy may improve fatigue.



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For PSC Partners
Community News,
summaries of the
Breakout Sessions
and of “Stress &
Resilience
Training,” please
continue with Part
II of the 2012
Summer issue of
The Duct ▶



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, provide education and support to PSC patients and their families, and promote awareness of PSC and organ donation.

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

To make a tax-deductible donation, please click on www.pscpartners.org/waystodonate.

Website

www.pscpartners.org

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