

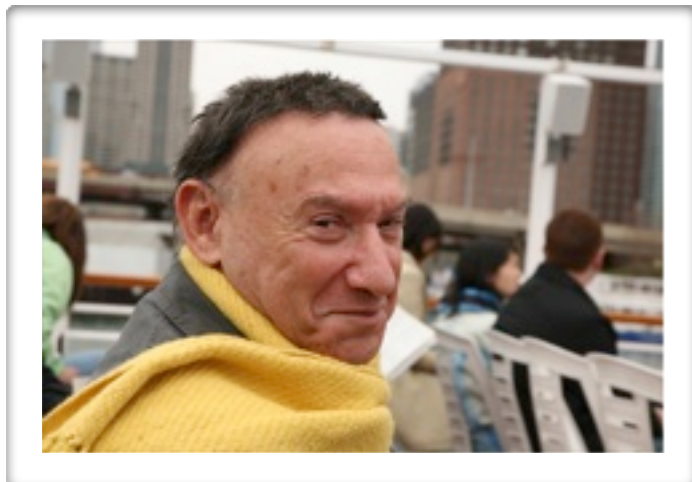
The Duct

Bringing you news on PSC



research, education, and support

PSC Partners Seeking a Cure Foundation Summer 2011 - II



We Lost a True Friend: Ivor Sweigler

Our entire global PSC community will be affected by the recent loss of Ivor Sweigler, Chairman of UK PSC-Support. Ivor inspired us all with his vivid intelligence, his passion in finding a cure for PSC and his efforts to help others affected by the disease.

When we were creating PSC Partners Seeking a Cure eight years ago, I contacted Ivor to introduce myself and to ask him many questions about his foundation. Ivor answered immediately with so much valuable information and a royal welcome. From that day on, he and I had a special friendship and a wonderful collaboration between our two organizations. Ivor came from London every year to attend our annual conferences and to give his brilliant presentations on PSC research progress in Europe. His loyalty to the PSC cause was unshakable.

Ivor's wonderfully quirky personality, mischievous twinkling eyes and devilish grin made him stand out in any crowd. He radiated such vitality, and every conversation with him was memorable. Ivor continues to be an inspiration to me. As one of our members wrote to me: "The world is a darker place without him."

Ricky Safer

A Tribute to Ivor

We all lost a friend... a loyal and true friend and a passionate advocate of PSCers. Even those among us who didn't know Ivor Sweigler, unwittingly have been touched by his immense impact on our PSC community. He tirelessly educated us, championed the PSC cause, and created a true partnership between the UK PSC group and all of us at PSC Partners. His intense commitment to the PSC community is permanently woven into the fabric and identity of our PSC family.

For those of us who met Ivor at our conferences - and he didn't miss a single one until this year - how could anyone forget his striking figure seated at the table closest to the speaker's podium, attentively listening and taking notes? Those notes would later turn into the conference summaries that have populated the special conference issues of our newsletters. He had the knack of giving a positive slant to the bleakest presentation.

For those of us who spoke to him at the conferences or heard him lecture, we experienced what Arne Myrabo described as his "katana-sharp wit," after the Japanese samurai sword known for its fine precision and for its complex yet delicate structure. He would not mince words, and with an intelligent and sharp quip, he would make everyone around him break into laughter.



Then there are those of us who knew him as a friend. His unwavering friendship, his sensitivity, and modesty made those of us who were his friends feel fortunate to have him in our lives. He knew precisely when to reach out, and would go the extra mile for those who sought his friendship. He touched hundreds of people all over the world, and he was a friend to all.

Ivor was one of the first people I met at my first conference in Jacksonville. Our friendship rapidly grew and in the past couple of years evolved within the space of this very newsletter. After the Chicago conference when he stayed with us, I would find him at dawn, sitting with earphones, paper and pen, fully focused on writing his conference summaries by hand. I took his papers and started typing his notes, and that started a partnership, with much laughter, overseas faxes, phone calls, and at the very end, he gave me a new friend, his wife Yumiko, the love of his life.

With the great void and sadness resulting from the loss of both our beloved newsletter editor and educator, Pat Bandy and Ivor Sweigler, we are left with two beautiful legacies. Their passion for improving the lives of PSCers, for being vocal and effective advocates of PSCers, and their superior intelligence and humanity will be important landmarks in the story of our path towards the cure.

Rachel Gomel

What YOU said about the Sacramento Conference

Thank you for providing us with this unique opportunity to learn, share, establish friendships with a remarkable network of inspiring people!

PSC Partners is the most important support group in the world for PSC. It plays an essential role.

It was a perfect conference, superbly organized, with super content, and super results.

Saturday Breakout Sessions

IBD Management in PSC

Dr. Sooraj Tejaswi, Assistant Clinical Professor, Division of Gastroenterology
Hepatology, University of California Davis

Dr. Tejaswi's PowerPoint presentation is accessible at <http://pscpartners.org/PSCConf11/PDFs/Tejaswi-IBD-PSC.pdf>

Introduction to IBD - Inflammatory Bowel Disease (IBD) is a chronic inflammatory condition of the gastrointestinal tract, probably caused by a combination of genetics, immunology, and extrinsic factors. There are two types of IBD: Ulcerative Colitis (UC) and Crohn's Disease (CD). UC is slightly more common than CD, in general. CD is rarely found in individuals with PSC.

The symptoms of IBD include abdominal pain, bloody diarrhea, and weight loss, which can also be caused by other factors, so diagnosis is a matter of differentiating IBD from other conditions which might cause those symptoms. The gold standard for diagnosis is a colonoscopy with biopsies.

UC looks different from CD. UC typically has a more uniform microcarpet of shallow ulcers. CD typically has small discreet and deep ulcers which may be surrounded by normal looking mucosa creating a cobblestone appearance. The pattern of colon involvement is also usually different. In CD, the rectum is almost always spared, and there are patches of normal tissue interspersed with diseased portions and the entire GI tract may be involved. There may be narrowing of the terminal ileum, fistulas, and perianal disease. In UC, 95 percent of patients have rectal involvement, and involvement is usually continuous rather than patchy.

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.

Treatment of UC & CD - The current medications available to treat IBD, from least toxic to most toxic, include 5-ASA medications including mesalamine or sulfasalazine, with brand names Asacol, Pentasa, Lialda; immunomodulators, e.g. Azathioprine, 6 MP [6-Mercaptopurine], Methotrexate; steroids; intravenous cyclosporine; and biologics (Remicade, Humira). The appropriate medication depends, among other things, on the severity of the disease, the distribution of the disease (location in the colon/GI tract), responsiveness to prior therapy, side effects, and patient compliance.

If 6-MP is used, liver function should be monitored every 2-6 months.

Surgical Management and Colorectal Cancer - UC is generally cured by removing the colon, whereas this surgery provides only symptomatic relief for CD. The preferred surgery for UC is a proctocolectomy (removal of the colon and the rectum) with ileal pouch anal anastomosis (IPAA), in which a J-pouch is formed from the terminal ileum. Another surgical option is to use the rectum to create the pouch (ileorectal anastomosis (IRA). This provides better pouch control and overall function, but leaves colorectal cells which are still susceptible to colorectal cancer (CRC).

CRC is one of the primary reasons a colectomy may be required, but it may also be used to treat UC that is non-responsive to medications. Surgery is not recommended to manage CD, but may be required to treat CRC, drain abscesses, or resect fistulas or strictures.

Other Complications - IBD is associated with a higher risk than average risk of osteoporosis, and anemia.

The use of sulfasalazine creates reversible sperm abnormalities in approximately 60 percent of men. Some of the surgical treatments, as well as pelvic inflammation in CD, may also decrease fertility. If pregnancy is achieved, and the disease remains in remission, pregnancy outcome is unchanged from that of the average population. If the disease does not remain in remission, there is an increased risk for both low birth weight and preterm delivery.

The risk of a child of a mother with IBD having a child with IBD is relatively low (1.6 percent for UC, and 5.2 percent for CD, but if both parents have IBD the risk increases to 36 percent).

The relationship between IBD and PSC - Seventy to 80 percent of people with PSC develop IBD but only 2 to 7.5 percent of people with IBD develop PSC. PSC is more strongly linked to UC (85-90 percent) than to CD (5-15 percent). Either can manifest first, although it is more common for IBD to manifest itself first because the average age of IBD onset is in the 20s and 30s, and the average age of PSC onset is in the 40s. Because IBD can be asymptomatic with PSC, it may have been present but not discovered until PSC diagnosis.

IBD in the presence of PSC - IBD typically is somewhat different when it coexists with PSC. There is usually continuous involvement of the colon (like UC), but the rectum is typically spared (unlike UC), and the terminal ileum is more likely to be involved (like CD). The disease is more likely to be mild, sometimes asymptomatic, but is more likely to carry with it a higher risk of colonic dysplasia and cancer.

The treatment of IBD is essentially the same, although IBD in the presence of PSC is more likely to be under treated because it is more likely to be asymptomatic. This may contribute to the higher risk of colorectal cancer.

PSC in the presence of IBD - The course of PSC does not appear to change based on whether IBD is present or not.

Colorectal cancer in the presence of IBD+PSC - The risk of colorectal cancer (CRC) increases dramatically when both PSC and IBD are present:

UC Duration	10 years	20 years	25 years
UC only	2%	5%	10%
UC + PSC	9%	31%	50%

The risk of CRC remains high, even if IBD remains in remission. There are more right sided cancers than average, perhaps because of an accumulation of toxins in that portion of the colon. Having a liver transplant does not appear to decrease the risk of CRC, and the risk is independent of gender. Symptoms which look like an IBD flare should be investigated for the possibility of CRC (e.g. weight loss, increased bleeding, and anemia).

Managing the risk of colorectal cancer -

Urso does not seem to prevent CRC, and it is not routinely recommended any more. Folic acid supplements may help, since they provide some protection in the general public. The cancer prevention role of 5ASA medications is unclear, but the side effects are so low that 5ASA medications should probably be used to treat IBD combined with PSC.

In IBD alone, colonoscopies are recommended every 1-2 years after living for 8-10 years with IBD (UC or Crohn's colitis). Once PSC is also diagnosed, annual colonoscopies are recommended. These colonoscopies should include a minimum of 32 biopsies, one every 10 cm in a spiral throughout the length of the colon. Because these still cover less than 1 percent of the surface area of the colon sampled, having an annual colonoscopy provides a better chance of catching dysplasia early. After a colectomy and the creation of an IRA, the pouch and rectal cells still need to be scoped annually because of the risk of cancer in the remaining rectal tissue.

PSC and impact on colectomy - Individuals with PSC who have had an IPAA have the same level of functionality of the pouch, and a similar quality of life as those without PSC. There is a higher risk of pouchitis (9-90 percent, as opposed to 7-47 percent), and the long-term mortality may be higher (probably

because of worsening PSC). Using the ileum to form the J-Pouch may contribute to the greater incidence of pouchitis in PSC patients because the ileum is more likely to be involved in IBD patients with PSC than those without. Multiple antibiotics may need to be tried if persistent or chronic pouchitis is a problem. The impact of removing the gall bladder on pouchitis is unclear.

Impact of IPAA on PSC - One small study indicated the level of PSC activity improved after IPAA, on average (50 percent improved versus 13 percent worsened, with 37 percent experiencing no change).

Impact of liver transplant on IBD - IBD should not be affected by a liver transplant, aside from the treatment benefit that might arise from the immunosuppressants used to prevent rejection. Most studies support this.

Summary - IBD-PSC may be a separate disease from IBD without PSC, having the unique features discussed above. Because of the likelihood that IBD will be silent in patients with PSC, every person diagnosed with PSC should be screened for IBD. If IBD is present, aggressive colon cancer surveillance should be undertaken, even if IBD remains asymptomatic.

Outlook for the near future: In the future we will see better colonoscopy imaging technology for early detection of colorectal cancer (chromendoscopy, the addition of pigments to better identify dysplastic tissue), blood tests to identify IBD patients at high risk for PSC, and drug trials aimed at preventing colorectal cancer.

Reported by Nancy Reeves

Transplantation and the MELD Score

Dr. Maurizio Bonacini, Department of Transplantation

Director, HIV-Liver Clinic California Pacific Medical Center (CPMC)

Associate Clinical Professor, UCSF

Dr. Bonacini's PowerPoint presentation is available at "[Pre-Transplantation and the MELD Score](#)".

Dr. Bonacini started by explaining that his main area of interest has been viral diseases and not PSC. He discussed the indications for liver transplant (OLT) in the U.S. and named hepatitis C as being the most common indication for OLT, with alcoholism, the number one killer, coming second (please see Dr. Bonacini's presentation for details).

The outcomes of OLT are most favorable for PSC and PBC (80 percent). He noted that the 2011 survival rate at California Pacific Medical Center (CPMC) was at 89.5 percent, versus the U.S. survival rate of 88.7 percent. Due to the numerous variables, he said that these numbers should be taken with a grain of salt.

He discussed data from UNOS describing the Model End-Stage Liver Disease (MELD score) currently used to allocate livers for OLT. MELD scores range from 6-40. Within this range, a MELD Score of 21-30 correlates to an average time of 128 days to OLT. A MELD score of 31-40 has an average wait time of 29 days. Status 1 patients (at risk of imminent death) have a mean time of 11 days to OLT.

He noted that having hepatocellular carcinoma and T1 tumors increase the MELD score. T1 tumors bring the MELD score to 20 while T2 tumors raise the MELD to 24. There has been an increase of OLT patients with malignant neoplasms. Before MELD, allocation of livers was based on length of time from listing. Since MELD, mortality rate has slightly decreased.

Pre-transplant, it is important to limit salt if ascites or edema are present. There may be a need for the patient to increase food intake if malnutrition is present, or to lose weight, if obese. Increasing vegetable protein intake is also helpful. Post-transplant patients need a low fat and low cholesterol diet. Adequate calcium and magnesium intake is essential.

Attention PSC Scientific/Medical Researchers:

Deadline for the next round of research grant applications is January 1, 2012. Our website has application guidance at this page:

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

PSC has a recurrence rate of 2-5 percent per year after OLT. The use of *Extended Criteria Donor* (reducing the stringent criteria for donor organs or increasing the age at which patients can receive a transplant) increases the risk of recurrence. Colectomy before or during OLT will decrease the risk of recurrence. The risk of colon cancer increases after OLT, and a yearly colonoscopy is recommended. The risk of IBD flares decreases after OLT.

Dr. Bonacini was asked about his thoughts on the ethics of medical tourism. He said that CPMC has had a history of complications treating foreign patients who had OLT outside the UNOS rules. They will no longer accept such cases. He also mentioned unethical practices such as paying donors to give parts of their livers. Living donor transplants do well with PSC patients and are an option at some hospitals.

Dr. Bonacini was asked about the possibility of quantifying pruritis as a MELD exception. Dr. Bonacini said that Regional Boards need to make exceptions. The question is complicated, he said, and cited research by a Yale researcher who invented a *scratchometer* to measure itching. He said that the earlier notion that bile acids in the blood were responsible for itching has been refuted.

Regarding the question of the MELD bias against PSC, Dr. Bonacini gave his own experience of a young PSC patient whose MELD score did not reflect the severity of her illness. He noted that the newest push for changing the MELD score is to include the sodium level into the equation, but he added that this change would not help PSCers. He encouraged attendees in this session and all PSCers to be vocal and to collectively press this issue with UNOS.

Reported by Kim Manfredi



**Would you be able to
help us with our
newsletter?**

*We need volunteers with experience in medical writing and newsletter editing. Come
and join our team!*

If you can help, please contact Ricky Safer at contactus@pscpartners.org

Interpreting Your Blood Tests

Dr. Christopher Bowlus, Associate Professor, Division of Gastroenterology and Hepatology, University of California Davis

Dr. Bowlus' handout is an important reference for PSCers who seek to understand their blood test results. Please find this document at "[Interpreting Your Blood Tests](#)"

The session was based on a discussion of the handout. Below are additional notes to complement Dr. Bowlus' handout.

- ALP represents ongoing injury to bile ducts. It is important to look for consistency of results. With diffuse disease, a PSC patient may have persistent chronic elevation. It is a concern when the level doesn't remain stable.
- ALT is much more specific to the liver. If ALT is elevated, it might signal autoimmune hepatitis.
- ALT and AST are liver injury markers. We like to think that lower levels of these two markers signify slower disease progression, but we do not have such evidence.
- AST is an indicator of muscle injury, and as such, is not as specific to the liver as is ALT.
- Bilirubin levels rise more quickly as obstruction increases.
- GGT is a more useful test in the pediatric population, as it can be indicative of PSC in that population.
- Platelet count (PLT) is one of the first indicators of portal hypertension. Hypertension is predicted by a drop in PLT and by an enlarged spleen.

Reported by Eve Jdrzejewska

The Role of ERCP in PSC

Dr. James Ostroff, Director of Pediatrics and Radiology, Professor of Medicine, University of California San Francisco

Dr. Ostroff's PowerPoint presentation is available at "[The Role of ERCP in PSC](#)".

Dr. Ostroff reviewed the process of evaluating suspected biliary obstruction. The general process is to first perform less invasive examinations, then to proceed to more invasive (and riskier) procedures.

Initially, magnetic resonance cholangiopancreatography (MRCP) is performed. An ultrasound (US) or computed tomography (CT scan) may be performed instead. If this initial examination appears to be normal, further evaluation to search for different reasons for obstruction follows. If the MRCP shows an obstruction,

an endoscopic retrograde cholangiopancreatography (ERCP) is performed. Surgery is rarely required at this point. The advantage of an ERCP is that 1) a narrowed bile duct can be dilated or stones can be removed, 2) cytology brushings can be taken to check for precancerous cells, and 3) bile ducts can be insufflated (expanded with dye) to make them more easily discernible.

In advanced cases where the common bile duct cannot be accessed by ERCP, it may be necessary to access the bile ducts externally via percutaneous transhepatic cholangiography (PTC). PTC frequently results in a biliary drain. PTC complications are different from those of ERCP. With PTC, infections and bleeding can occur while ERCP can result in pancreatitis. PTC is not the test of choice for most endoscopists.

Dr. Ostroff noted that the MRCP procedure has improved dramatically in the last decade. Currently for diagnostic purposes, MRCP is almost as good as ERCP. Though ERCP still has several advantages over MRCP, it is dangerous and can cause problems. One of the more common complications of ERCP is pancreatitis (inflammation of the pancreas). Pancreatitis is caused by the proximity of the ending of the common bile duct to the ending of the pancreatic duct. Even though the endoscopist does not go into the pancreas, any inflammation or “bruising” can cause the pancreatic duct to swell itself closed. Most pancreatitis instances are self-limiting.

The key points to a safe ERCP are having, 1) a trained and experienced staff 2) an adequate back-up 3) a complete medical resuscitation of the patient 4) slow and methodical sedation 5) a team that is not rushed 6) liberal use of the anesthesiologist in charge.

Many tools are available for performing ERCPs: dilating balloons, cell-sampling brushes, stone removal tools, etc. He noted that UCSF very seldom uses stents, and then, only in special cases, as a bridge to health or transplant. If used, they are removed as soon as possible.

In summary, diagnostic ERCPs are used to make a PSC diagnosis, stage the disease and obtain cytology samples to exclude cholangiocarcinoma. Therapeutic ERCPs (dilations) are part of a multidisciplinary approach with an established clear endpoint, and can serve as a “bridge” to liver transplantation.

Reported by Arne Myrabo





Ask a Chef: Natural Cuisine for Healthy Cooking

Russell Michel, Executive Chef, Sheraton Grand, Sacramento
Celebrity Chef Winner 2010, Fox 40 Live Ask a Chef

Chef Russell Michel's PowerPoint presentation is available at <http://www.pscpartners.org/PSCConf11/PDFs/Michel-Eat%20Well,%20Live%20Well-2.pdf>

Eat Well, Live Well. Those simple words served as the starting point for Chef Russell Michel's presentation, *Ask a Chef*, during our Saturday breakout session. Could it be that simple? Can we, as PSCers, eat well and live well? It seems, perhaps, the answer could be yes. Chef Michel had been feeding us all weekend. We'd had delicious pizzas and sumptuous granola, delectable cookies and superb salads. We were all abuzz and ready and eager to learn how we could use food to help ourselves heal and mend while enjoying recipes that even non-liver patients crave.

Chef Russell began his presentation by describing the difference between organic and commodity, a distinction he assured us can and does make a difference in our physical health. Commodity-raised items are generally less healthful, fed inferior quality ingredients which, in turn, get passed into our systems, and may contain hormones, antibiotics or pesticide ingredients which we then are unwittingly delivering into our systems. Organic items, however, pose few if any of these risks, particularly if you know exactly where the food comes from.

He also shared with us the story of his brother-in-law, Robert. Robert was a 46-year-old man in need of a liver transplant. He wasn't a PSCer, but his liver was failing. Chef Russell asked if he could step in and try helping him through diet and nutritional changes. His brother-in-law said yes and, within the short time span of just five months, doctors were shocked to see that positive changes had occurred. But why?

Well, according to Chef Russell, the answer is relatively simple (all things considered). He presented us with a diagram that helps explain his views (PowerPoint Slide 4). In the slide, he shows what our real pyramid should look like, the ways we can utilize food for not only nourishment of mind and soul, but also body. By focusing on healthful, local ingredients, keeping well hydrated and eating a balanced diet, along with some form of daily physical activity, we, too, may be able to reap significant changes in our health status. His top suggestions included fruits and veggies, as much as you can get in, but ideally at least 6-9 cups a day, whole grains, lean proteins, a moderate level of heart healthy fats, such as olive oil, and limited sugar consumption (alternatives to traditional sugar presented in PowerPoint slides). Meat isn't off limits, but it should be used in moderation and chosen with care, grass fed over grain fed, fish that come from clean waters. And for those of us who are vegetarian, Chef Russell also reminded us that protein doesn't only come in meat form. In fact, he urged even the carnivores among us to try to ingest more plant-and dairy-based proteins, citing the fact that they help fatigued muscles, they're lower in saturated fats and they are quickly digested by our bodies.

Once we covered foods in general, we moved on to what we should be buying if possible. In season fruits and vegetables top the list. Chef Russell advises against frozen and suggests instead canning your own fruits and veggies when they're available to ensure you have access to the same level of healthy ingredients even in a food's off-season. He also urges purchasing local labels and organic produce when possible. This way, you're not only stimulating your local economy, you're also able to have a connection to the food, to know where it's grown and trust what's in it. If you don't live in farm country, not to worry. There are Farmer's Markets all over the United States and all over the world. There are local organic farms that host CSAs (crop shares where they send you the freshest of what's available on their farms at any given time) and, last but not least, the organic section of your grocery store.

So, you may be wondering what exactly happened to Chef Russell's brother in law, Robert. Well, five short months after incorporating the chef's dietary changes into his life, Robert's MELD dropped from 29 to 17. When asked if he was surprised, Chef Russell smiled. It only makes sense, he said, that our bodies would respond in a positive way to being treated with care. Where food is fuel and fuel is energy, how could we go wrong paying more attention to what we put into our systems? For Chef Russell, he's not naive and he understands that diet alone may not cure PSC, but as he states, it's his job to keep us "staying well while seeking a cure." As for me, I don't think we can ask much more than that. :)

For more suggestions and ideas to stay well while seeking a cure, please check out the PowerPoint presentation of Chef Russell Michel here on our website and for some of Chef Michel's recipes, check out our "Diet/Nutrition/UC/Gluten Free/Recipe Share, etc." document on PSC Partners Seeking a Cure, FaceBook.

Reported by Sandi Pearlman

Liver Transplantation for PSC

Dr. John P. McVicar, Professor of Clinical Surgery

University of California Davis

Dr. McVicar's PowerPoint presentation is available at ["Liver Transplantation for PSC"](#)

Surgeon John P. McVicar, M.D., led a productive interactive breakout session focused on liver transplantation as a treatment for PSC. Dr. McVicar is a Professor of Clinical Surgery with more than two decades experience in abdominal transplant surgery prior to the discontinuation of liver transplantation at UC Davis Medical Center. Dr. McVicar provided a very thorough and clear summary of the transplant procedure. He provided an overview of the history of liver transplantation in the United States, beginning with the pioneering work of Dr. Tom Starzl, and continuing with an overview of the complexities of the procedures and the hurdles to overcome before achieving the current high success rates. The overall one-year survival rates for liver transplantation have now been approaching 90 percent for the past decade, with success for PSC patients being among the highest.

One of the points Dr. McVicar made early in his presentation was that not every PSC patient will necessarily need a liver transplant. Although the treatments are limited at present, he notes that as individuals are diagnosed earlier and control of symptoms improves, an increasing number of PSC patients will be able to live with the illness for quite some time before liver transplantation becomes necessary. In presenting liver transplantation, he noted that it is a treatment, not a cure. As such, the recommendation of when to proceed with transplantation is made when the risks of underlying liver disease become greater than the risks associated with transplantation. Key markers of liver decompensation discussed include uncontrollable ascites, pronounced jaundice, bleeding varices, and hepatic encephalopathy. However, he notes that physicians may recommend transplantation in the presence of recurrent cholangitis episodes, severe itching, or a history of certain types of primary cancers.

When discussing the advancement of liver transplantation success, Dr. McVicar highlights numerous refinements in surgical techniques, improved immunosuppression regimens, better organ preservation, and improved patient selection and management. Despite these advances, Dr. McVicar stressed that there remain many risks associated with the procedure, particularly during and immediately following the surgery. Individuals with MELD scores above 30 often are at greatest risk.

Before outlining the specific risks, Dr. McVicar provided an overview of the transplant procedure in relatively understandable, non-technical language. He noted that one of the most technically demanding parts of traditional cadaveric transplantation is removing the damaged liver. The risk of bleeding during this part of the procedure is relatively high, particularly for individuals with extensive adhesions (often from prior abdominal surgeries) or for those with marked liver decompensation. Prior to beginning this part of the procedure, most surgeons place patients on bypass to ensure continuous flow of the blood and to maintain body temperature. Once a patient is on bypass, the process of carefully separating the damaged liver from other tissues is completed, ending with separation from the blood supply and removal of the liver. The new liver is then placed in the abdomen and connections made to the primary blood vessels and the bile duct. Specifically, there are five connections made between the new liver and the transplant recipient: the vena cava (the large vein in the abdomen that returns blood to the heart) both (1) above and (2) below the liver, (3) the portal vein (taking blood away from the GI tract to the liver), (4) the hepatic artery (the main blood supply to the liver), and (5) the bile duct. The surgeon typically attaches the bile duct exit from the liver directly to the small bowel while eliminating the bile ducts outside of the liver in order to reduce the risk of scars to the bile ducts and reduce the risk of bile leakage.

Dr. McVicar noted that the surgical procedures are largely the same for live donor transplant recipients, though making the connections to the blood vessels is much more technically complex. This is because the blood vessel segments on the donated organ are much smaller. In addition, the vena cava connections are significantly different since both the donor and recipient require an intact vena cava.

Once Dr. McVicar had summarized the procedure, he discussed some of the specific risks and potential complications. The most pronounced risk during surgery is bleeding, as already mentioned. He noted that heart attack due to the stress of the surgery is a potential risk, but that this is quite unlikely with proper patient screening and treatment of underlying heart disease prior to the transplant. Once the transplant is complete, there is a risk of a clot forming in the hepatic artery (hepatic artery thrombosis). Although this is relatively infrequent, formation of such a clot often results in an immediate need for a re-transplant. The first 24 hours present the greatest risk. Although risk for such a clot remains elevated

for the first week or so following transplant, the first 24 hours are the most critical. A less common risk is primary non-function of the new liver. Although this is quite rare, it is another indication for immediate re-transplantation.

The more common complications are quite treatable. For instance, other surgical or technical risks that can cause problems either shortly after surgery or on a delayed basis include bile leaks and bile duct strictures due to scarring. Other types of complications tend to be medical in nature, meaning that they are usually treated with medications. Rejection occurs in 10-20% of patients. Dr. McVicar emphasized that this is generally of little concern as long as it is caught and treated early. When caught early, rejection episodes often require only adjustment in medications or brief stays in the hospital to monitor these changes.

Due to immunosuppression, another common complication is infection. Infections can be viral, bacterial, or fungal. As with rejection, most infections are treatable. Although the risk of infections is highest in the months after surgery, infection may also be a chronic risk post-transplant, particularly for those requiring higher doses of immunosuppressants.

The last complication Dr. McVicar addressed is the risk of recurrence of PSC. Since the causes of PSC are not well understood, it is not surprising that recurrence of the disease is not well understood either. Dr. McVicar noted that the best estimates at present are that 20 percent of PSC patients are diagnosed with recurrent PSC within the first 10 years of transplant, meaning that 80 percent of PSC patients do *not* experience recurrent PSC.

One of the issues that aroused the greatest interest in the session was the discussion of living donor liver transplantation (LDLT). Dr. McVicar noted that LDLT remains an important approach to liver transplantation despite the decrease in the number of LDLT procedures performed annually since the procedure frequency peaked in 2005. Although the procedure accounts for only about 5 percent of the total liver transplants performed in the United States each year, LDLT accounts for anywhere from 10-20 percent of transplants for PSC patients, depending on the year. As an example, LDLT accounts for 219 out of 6101 transplants in 2009 and 282 of 6009 transplants in 2010, that is, 3.6 percent and 4.7 percent, respectively. However, it represented 29 out of 261 transplants for PSC patients in 2009 and 54 out of 256 transplants for PSC patients, that is, 11.1 percent and 21.1 percent, respectively.

However, not all centers perform LDLT, and not all centers perform them equally well. Such procedures have only been in place for a little over a decade for adult-to-adult transplantation. With the relatively low frequency of LDLT, the novelty of the procedures, and the extreme technical demands placed on the surgeon (e.g., connecting the hepatic artery to a donor blood vessel measuring around 1/10th of an inch), many centers do not have surgeons capable of performing the procedures. Even the larger centers perform only around ten LDLT per year. Dr. McVicar noted that this procedure remains a viable, effective option for many individuals, particularly for PSC, given its slow progression and chronicity, allowing scheduling of the procedure. He noted that this is particularly valuable in those parts of the country where cadaveric transplants are occurring only at very high MELD scores. However, he encouraged careful research prior to considering LDLT as an option, noting that if he were a patient, that he would seek out one of the centers performing the highest number of the procedure. This information is available through the United Network for Organ Sharing website www.unos.org.

In addition to the information described above, Dr. McVicar discussed several issues related to the donor. For instance, he noted that donors need to be quite healthy, nonsmokers, and well positioned financially, psychologically, and socially, to recover. Although most donors recover quite well, complications do occur. The process of donating a portion of the liver from an adult to a child is relatively straightforward, but adult-to-adult procedures are quite complex. Although he stated he feels adult-to-adult procedures play a very important role in liver transplantation for PSC, he also noted that some hepatologists say behind closed doors that they would not consider donating a portion of their livers to a family member.

After discussing LDLT, Dr. McVicar fielded many questions from the conference participants. The first set of questions related to future direction in transplantation including xeno-transplantation, creating livers from stem cells, and using stem cell generated liver cells in artificial livers. With regard to xeno-transplantation (i.e., using organs from another animal), Dr. McVicar noted that there was early hope for using pigs, but that a protein present in pigs causes clotting, making this implausible without genetically breeding animals without this protein. Rejection rates are also quite high with any form of xeno-transplantation. He noted that there was an effort to breed a genetically altered pig to eliminate this protein and reduce other problems with rejection, but that progress is slow and that they are not anywhere near a phase two trial at this point.

With regard to the use of stem cells, significant progress has been made. It is feasible at this point to create liver cells. However, creating the physiological “scaffolding” to guide the cells into a structure that will lead to a usable organ remains in its early stages. He contrasted this with the rapid regeneration of liver cells and structure in a LDLT. In a LDLT, the structure of each portion of the liver remains intact, allowing for rapid reproduction of liver cells to take on the needed volume. This often occurs within a matter of weeks. However, stem cell generated liver cells have no structure upon which to grow. As technologies advance, it may someday be possible to generate an organ from a patient’s own cells. At this point, however, a more feasible solution based on stem cells or one’s own cells is on the horizon and includes the possibility of artificial liver machines. These could filter someone’s blood in much the way dialysis machines work. In contrast to kidney dialysis, however, liver machines would require living liver cells due to the complexity of the liver’s functions. Dr. McVicar emphasized that he is not an expert in this field, but he shared his enthusiasm with the group over the long-term promise of some of these technologies.

Another question raised concerned the risk for bleeding. Several in the group with low platelet counts were interested in whether this greatly increased the risk for bleeding. Dr. McVicar offered a very hopeful outlook for these patients, noting that he generally was not concerned about low platelet counts. Rather, excessive adhesions in the abdomen around the liver were a greater concern. When this raised concerns from others in the group with a history of extensive abdominal surgery, Dr. McVicar noted that previous surgeries did increase risk but that he found that he had trouble predicting from someone’s history who would bleed more than others. Rather, he noted that an extensive history of prior surgeries was primarily just an annoyance to more experienced surgeons because it would mean more hours on the table. Dr. McVicar also discussed other concerns regarding potential complications. With good humor and detailed information regarding how surgeons could prepare for and address

complications as they arise, most in the group seemed to find increasing hope for positive outcomes from the transplant process.

Dr. McVicar concluded the discussion by addressing questions regarding the MELD system. The most frequent questions raised regarded potential exceptions or “extra points” to the MELD system. He noted that there are currently approximately a dozen different exception criteria and that they vary by region. Although several were mentioned, the most common among PSC patients with whom he has worked has been points for recurrent cholangitis episodes.

In summary, Dr. McVicar provided a thorough, informative, and engaging discussion about transplant surgery as one treatment for end stage liver disease resulting from PSC. His presentation provided a discussion of the risks and challenges to the surgery in a realistic, informative fashion yet provided participants with significant hope. Hope was evident in the positive outcomes seen with the current state of transplant surgeries as well as the potential for alternatives to transplantation many years in the future.

Reported by Philip Burke

Coping for PSCers: Making Your Life More than PSC

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

Dr. Burke's PowerPoint presentation and his handouts are available at "[Coping for PSCers: Making Your Life More than PSC](#)". Attendees of this session found the presentation and the handouts to be valuable

Philip Burke gave a comprehensive presentation on coping with PSC for patients. He began by talking about stress that is prevalent at all times in our life, independent of PSC. Added to those daily stresses, stress resulting from having the wrong set of healthcare providers, for example, might go as far as to cause patients to question themselves and result in distress.

It is important for each individual PSCer to find strength and appropriate techniques to make life greater than the illness. Each PSCer has to deal with the different ways in which PSC could take control of his/her life. For some, PSC encompasses their whole life while for others, at the onset, PSC does not consume them, but down the road, causes problems.

Some PSCers are able to remain positive, appreciate life and keep things in perspective, and some others go even further and attribute greater meaning to PSC and consider PSC a reason to make life more meaningful. The human body is built to respond to short-term physical and psychological demands. This type of “acute stress” leads to rapid thinking that is based on past experience and that narrows our focus and attention. Conversely, this kind of stress inhibits creative thinking, restricts our range

of emotion, and impairs social judgment. PSC patients cannot rely solely on short-term thinking as if preparing for danger because the demands of PSC are ongoing rather than acute and include physiological changes, unpredictability, and difficulty in finding ways to focus on action.

With the challenges of living with long-term stressors come numerous styles of coping. These can be divided into two major categories, *engagement* and *disengagement*. In the *engagement* category, people are problem-focused, display a fighting spirit, and are socially oriented. This method of coping involves logical thinking that enables the person to focus on emotions without becoming overwhelmed. With a disengaged style of coping, a person focuses on emotions, blames others, can resort to substance abuse, wishful thinking, and isolation.

Associated with these strategies are certain myths that give preference to one style over another. For example, one common myth is to view avoidance and denial as being unhealthy; however, at times, fully engaging one's emotions could be dangerous and overwhelming, and avoidance could serve as an adaptation mechanism for someone facing a new situation. Another myth is that problem-oriented coping always works best. This holds true when the problem is controllable and solvable, but it is important, at times, for the patient to accept what cannot be changed. A third myth is the common notion that working through emotions is always valuable; however, at times, inwardly focusing on emotions may lead to disengagement. The fourth myth Dr. Burke spoke about describes self-pity as a harmful practice. He debunked this myth by explaining that the occasional "pity party" is an expression of honesty and might serve to reach a state of acceptance of oneself and bring a realization of the positive elements present in our lives.

Patients may encounter various sets of problems throughout the course of their disease, and these can range from having issues with healthcare providers to suffering from depression. All PSCers have one thing in common, and that is their relationship with healthcare providers. These relationships work well when patients are coping well because these patients are able to accept that healthcare providers, like everyone else, are flawed human beings trying to help their patients to the best of their ability. These patients are also able to forgive those who cannot actually help them.

A handout on assertiveness focused on helping patients build assertiveness skills. He emphasized that patients are at the center of their healthcare and must be their own advocates. He suggested that patients have specific questions and ideas ready for each appointment. It would also be helpful to bring along an advocate to take notes of the physician-patient exchanges.

Dr. Burke addressed specific PSC challenges such as fatigue, sleep, depression, relationships, and sexual well-being. He explained that the inflammatory process could possibly bring about changes in cortisol, melatonin and other mood-affecting hormones, sleep-wake cycle, and certain aspects of life. With sleep problems may come a change in mental capacities. He provided a list of suggestions for coping with sleep disturbances. He recommended practicing acceptance, having balanced nutrition, staying hydrated, taking brief naps, moderating caffeine intake, minimizing light and sound, and setting worries aside.

Lastly, Dr. Burke discussed depression. He explained that the symptoms of depression, PSC and IBD were similar, that depression was not a weakness, and that one could be depressed without being sad.

Of those experiencing depression, he said that 61 percent presented with physical symptoms and not with sadness. For patients with depression, help is available through medication, cognitive and cognitive-behavioral therapies. In some cases, relying on friends may be counter-productive. He advises PSC patients to stay proactive and active, address what they think-feel-do, reduce isolation and seek support, build mastery, talk with healthcare providers, and maybe consider other professional help.

Reported by Julianne Vasichek

Pediatric PSC and Potential Therapies

Dr. Philip Rosenthal, Professor of Pediatrics and Surgery, Director Pediatric Liver Transplant Program, Director Pediatric Hepatology

University of California San Francisco

Dr. Rosenthal's PowerPoint presentation is available at "[Pediatric PSC and Potential Therapies](#)"

Dr. Rosenthal explained that the disease presents itself differently in children than in adults and that prevalence is different for adults across genders.

The diagnosis of PSC is made differently with children, as blood tests, and particularly liver function tests (LFT), are not frequently prescribed for this age group. With children, PSC is usually discovered through follow-up testing for ulcerative colitis (UC) and Crohn's Disease (CD). With children, PSC does not present with bile duct injury. In adults, however, raised LFTs are the initial source of investigation.

In children alkaline phosphatase (ALP) is not indicative of PSC because ALP levels are also affected by bone growth and can be misleading. For diagnosis, MRCP is the best and least invasive way of imaging the bile ducts. Gamma glutamyl transpeptidase (GGT) is often elevated in children with PSC, but in adults elevated GGT levels are not indicative of PSC.

Children, unlike adults, often present with autoimmune hepatitis (AIH) overlap syndrome (1/3 of children have PSC and AIH overlap). These cases respond well to immunosuppressive drugs (Imuran) and steroids. It is believed that AIH overlap may be an early stage of PSC in children. Though the progression of the AIH component of the illness can be controlled, the progression of bile duct injury can continue independently.

In treating overlap syndrome, it is important to limit exposure to steroids due to their negative impact on growth, their potential for bone injury, and their other side effects. If blood tests and biopsies are normal after one or two years, then an attempt is made to wean the child off steroids.

Ursodiol in the suggested range of 10-20 mg/kg/d is another treatment used for PSC. As the child grows, the dosage is adjusted; ; however, if blood tests are within the normal range, the dosage is not

increased. Currently, STOPSC is studying the effect of Ursodiol on children.

A recent study using oral Vancomycin with 14 children had positive results. Ten of the fourteen children with no cirrhosis normalized and 4 with cirrhosis improved. Improvement was noted both in clinical symptoms as in LFTs. The concern with this study is that the sample size was small and the trial was not randomized (children had both PSC and IBD). With Vancomycin, there is a risk of developing Vancomycin-resistant enterococci (super bug), which can be life-threatening. Given the good results of this small study, larger randomized clinical trials to assess the use of Vancomycin for PSC in children as in adults are warranted.

Itching, a common symptom with pediatric PSC patients, is treated with Ursodiol, Rifampicin (may lose its effect with long-term usage), and Cholestyramine (Questran); the latter may be difficult with children, as Questran requires large fluid intake.

Bile duct cancer (cholangiocarcinoma) is exceedingly rare in children, and liver transplantation has very good outcomes. As Dr. Rosenthal stated in his final slide in his presentation, the important messages to take home are that PSC is rare in children; no controlled clinical trials were performed in children with PSC; STOPSC is now attempting to accomplish the much-needed multi-center collaborative research to find out more about pediatric PSC.

Reported by Eve Jedrzejewska

A huge thank you to the following individuals who are personally sponsoring a research project, either through a personal donation or through their fundraising efforts:

Anonymous donor

Rachel and Abe Gomel

Hoops4Healing

Scott and Susan Malat

David and Ros Parry

Daniel Schachter

Ken and Patty Shepherd

Craig and Ali Wiele

Sheldon and Robin Wohl

Screening and Treatments for Cholangiocarcinoma

Dr. Carlo Selmi, Assistant Professor, Division of Rheumatology, Allergy and Clinical Immunology, Department of Internal Medicine, University of California Davis

Department of Translational Medicine, University of Milan

Dr. Selmi's PowerPoint presentation is available at "[Screening and Treatments for Cholangiocarcinoma](#)"

What is Cholangiocarcinoma? Cholangiocarcinoma (CCA) is an aberrant proliferation of bile duct epithelial cells which are the cells covering the surface of the bile duct. They multiply much more quickly than they are intended to multiply, and become malignant. CCA is the second most common cancer originating in the liver.

CCA can be found at any level of the biliary tree. When CCA is found in the bile ducts inside the liver, it is called *intrahepatic CCA*. These tumors account for 25-50 percent of all CCA. When CCA is found in the bile ducts outside of the liver it is called *extrahepatic CCA*. These account for 50-75 percent of all CCA. The remaining are found where the right and left hepatic bile ducts join together, at the entry into the liver. These are known as *Hilar* or *Klatskin* tumors.

Wherever the tumors are found, they can line the bile duct, causing constriction, or present as masses which grow into and out of a focal point on a bile duct.

An intrahepatic tumor usually begins as a mass in the small bile ducts. Intrahepatic tumors are harder to find in the early stages. Their location in the smaller bile ducts means that they tend to create less dramatic symptoms and are often only found by accident. Typical symptoms include fatigue, malaise, abdominal fullness or discomfort, right upper abdominal mass, and weight loss.

Extrahepatic tumors, on the other hand, tend to narrow the large extrahepatic bile ducts and create much more obvious symptoms. Ninety percent of people with extrahepatic tumors present with severe jaundice and mild pain. Other symptoms of extrahepatic tumors include clay colored stools, pruritis, dark urine, abdominal pain, and weight loss.

The Epidemiology of CCA - In the general population, CCA is rare, occurring in .85 people per 100,000 people per year. There are approximately 5,000 new cases each year. Out of 169 people diagnosed with PSC in the Kaiser Permanente database, 7 developed CCA over a 5-year period.

New diagnoses of CCA are increasing slightly, primarily because more intrahepatic CCA is discovered. Diagnoses of extrahepatic CCA are decreasing slightly. Increased frequency and sensitivity of imaging techniques likely account for the increase in diagnosis of intrahepatic CCA. This is particularly true in the PSC population, which is subjected to more frequent imaging than the general population. The location of the PSC involvement may affect the diagnosis, because it is hard to distinguish benign from malignant strictures. Taking brushings while attempting to open apparently benign strictures may lead to the discovery of CCA.

Most patients with PSC will not get CCA, and CCA generally spares individuals with small duct PSC. The cumulative lifetime incidence of CCA is between 9 and 20 percent. The risk of CCA is between .5 percent and 1.5 percent per year. The median age of diagnosis of PSC patients with CCA is between 30 and 50, as compared to 65 for non-PSC patients. Neither the presence of UC, nor the removal of the colon, changes the risk of CCA.

Only ten percent of CCA diagnoses are associated with recognized risk factors. Anything which causes chronic inflammation, such as PSC, is associated with an increased risk of CCA. Established risk factors for CCA include liver fluke infestation, choledocal cysts, hepatolithiasis, thorotrast infusion, and PSC. CCA is also weakly linked to alcohol intake, tobacco use (the opposite of UC), chronic viral hepatitis without cirrhosis, and obesity.

Screening for Diagnosis of CCA - Early diagnosis is crucial to a positive outcome. The optimal frequency of screening for CCA in PSC patients is unclear, but screening on diagnosis and regular later screening are recommended.

For asymptomatic patients, Dr. Selmi does the following: an ultrasound (US) every 9-12 months (6 months if cirrhosis is present) and standard liver bloodwork (not including CEA or CA 19-9) every 6 months). Some physicians also use additional imaging techniques: CT, MRCP, ERCP, PET, endoscopic ultrasonography (EUS), and intraluminal cholangioscopy (IC). These additional imaging techniques carry a higher price tag, and not everyone agrees that the cost of screening PSC patients annually via MRI, PET, or ERCP is justified by the few cancers detected.

Some physicians use serum markers for screening. Marker sensitivity and specificity of these markers make them not well suited for use as routine screening tools because they generate both too many false positives and false negatives. *Specificity* measures the chance that a positive test means you have the disease. One hundred percent (100%) specificity means there will be no false positives. *Sensitivity* measures the chance that you will test positive if you actually have the disease. One hundred percent sensitivity means there will be no false negatives.

With diseases involving chronic or acute cholestasis, such as PSC, the specificity of CA 19-9 is far lower than it is in the general population. Using data taken from the general population, the carbohydrate antigen (CA 19-9) has a sensitivity of 94 percent at ≤ 40 u/ml, and CA 19-9 has a specificity of 100 percent at > 129 u/ml. Dr. Selmi indicated that numbers significantly higher than 129 u/ml, though, can be caused by cholangitis, even when CCA is not present.

Carcinoembryonic antigen (CEA) is also sometimes used as a screening tool, with similar challenges. Some studies indicate a CEA > 5 $\mu\text{g/l}$ has a specificity of 86 percent and a sensitivity of 55 percent in suspected CCA. CEA is also sometimes used to evaluate suspected colon cancer.

Cytological evaluation of brushings, biliary cell samples obtained when dilating strictures or during an ERCP, is another screening tool used for CCA. The sensitivity of the tests used on these cells is below 50 percent (lots of false negatives). This is because brushings collect isolated cells, and less information can be gained than from studying the tissue (histology) that would be obtained in a biopsy. The sensitivity does improve, however, with repeated sampling. The specificity is between 97 percent and 100 percent (almost no false positives). FISH (fluorescence in situ hybridization) techniques, used to detect and localize the presence or absence of specific DNA sequences on chromosomes, may be added to the cytological

evaluation. It is unclear whether FISH techniques increase the chance of finding CCA, but the degree of suspicion is likely related to increased success.

Dr. Selmi is not in favor of using either serum marker for routine screening (screening in the absence of a specific suspicion of CCA) because of poor sensitivity and specificity.

A suspicion of CCA would arise because a lesion is detected using an imaging test, a change in symptoms, or persistent jaundice, persistent pruritis, weight loss, abdominal pain, or rapidly increasing serum bilirubin. The initial evaluation would include preliminary imaging. If a lesion is detected on ultrasound, but no other symptoms are present, Dr. Selmi would follow the ultrasound with an MRCP, then evaluate the next steps based on what was found. In the absence of symptoms, an ERCP with brushings would not be his first step. Serum markers might be added at this stage to rule CCA in, or out, as a cause of whatever it was that raised the suspicion.

If a liver lesion is detected from the MRCP, an ultrasound or CT guided biopsy is recommended with a histological examination of the biopsy. EUS guided fine-needle biopsy has 100 percent specificity and 91 percent sensitivity in cases of suspected CCA. The risk of spreading the cancer is generally lower than the need for a firm diagnosis. In addition, the lesion may be secondary to cancer elsewhere, which can generally be determined from a biopsy.

Because the symptoms of extrahepatic CCA mimic PSC, a benign stricture, or end stage liver disease, diagnosis is notoriously challenging.

If a new stricture is being dilated, brushings should be taken to evaluate for CCA. If the brushing is negative, an EUS is recommended to look for masses. Over half of CCA diagnoses are made at the same time, or within a year of diagnosis of PSC. PSC has likely been present for a number of years, and the CCA may have created the symptoms which led to the diagnosis of PSC. Even if the brushings are negative, the stricture should continue to be monitored.

Treatments - CCA is significantly more likely to be caught early than it used to be because of more regular imaging and brushing cytology. In the early stage, tumors are often resectable. If the lesion can be completely resected, there is no lymph node involvement, and no metastasis, the survival rate is 50-60 percent at 5 years. Resection may not be possible for patients with PSC because the liver may be too damaged to permit the remainder to do all of the work the liver needs to do.

Transplant is also now a potential cure for CCA patients, particularly with hilar (or perihilar) tumors. Transplants are being done at Mayo, UCSF and other locations. CCA used to eliminate patients from receiving a liver transplant, but more recently it has been discovered that patients with CCA, diagnosed from the excised liver, did well following transplant. Approximately 75-80 percent of individuals treated by transplantation are CCA free five years post transplant. A live donor may be used in a transplant to treat CCA.

If surgery is not an option, the remaining treatment options include chemotherapy, radiotherapy, and endoscopic ballooning or stenting to relieve the symptoms caused by the strictures.

Conclusions - CCA is a rare cancer, and most people with PSC will not develop it. If they do, more frequent imaging is catching lesions earlier, and treatment is most effective if CCA is caught early, so the chances for survival are improved.

Reported by Nancy Reeves

Practical Considerations Before Transplant

Panel of Post-Transplant PSC Patients and Caregivers

Moderator: Joanne Grieme

Panelists: David Abbinanti, Alison Collins, JoAnn Collins,

Tom Butler, Aubrey Goldstein

Approximately thirty people attended the session. Four among them are on the waiting list for a transplant.

Joanne welcomed everyone and asked the panelists to introduce themselves, tell their transplant story and offer a piece of advice.

Aubrey had his transplant on May 10, 1998. He had liver disease for 13 years before they diagnosed PSC. Things have gone pretty smoothly since transplant.

JoAnn, is the mom of Alison who had her transplant on November 29, 2010. JoAnn was glad she had been told that the surgery would likely go smoothly but that there could be complications following the transplant. This was the case with Alison, and for three months they had to live away from their own city.

Alison was diagnosed with ulcerative colitis at age 5 and PSC at age 15. She received part of her sister's liver. She encouraged PSCers to eat frequently in order to get enough nutrition and stay strong.

Joanne is the mom of Todd, who is now 25. She was quick to say that not everyone has the same difficulties as Todd has! He was diagnosed with PSC at 15 and received part of his brother's liver at 17, but shortly after, received a cadaver liver due to surgical (not liver) complications from his first transplant. His PSC recurred one and a half years after transplant. He had a third transplant, and is now doing well! Joanne told the group to expect a six-month recovery period after transplant.

David was diagnosed in May 2006 and received part of his sister's liver in August 2009. He emphasized the importance of remaining positive no matter what is thrown at you. He had hernia surgery in December but is doing well.

Tom was diagnosed in 1999 and in December 2008, had a living donor transplant thanks to the generosity of a member of his church. He recommended doubling the time that doctors say everything will take, and ensuring that your financial records are in order. He also emphasized the importance of recognizing that caregivers need support, too.

Following the introductions, a number of questions were raised regarding living donors. Although size is an issue, David indicated that he was able to receive 68 percent of his sister's liver. All the donors are doing well. It was also confirmed that donors needed to have the same blood type but that Rh factor is not an issue. Although transplant centers can have different policies, it is generally expected that the donor have a reasonably close relationship with the recipient.

The post-transplant hospital stay varied greatly, between 8 and 17 days, with some needing re-admission soon after due to complications. But Alison and Aubrey both emphasized that they felt better immediately following surgery, despite the surgical pain.

The recipients also said that their energy level improved after transplant, and that the medications were manageable. They said that once they started to feel better, it was easy to forget to take their pills. It is, however, greatly important to take the prescribed medication regularly and at the same time each day.

The recipients all agreed that the wait for a transplant was very difficult, both mentally and physically. They said it was important to keep spirits up and remain as active as possible. They recommended that while waiting, it would be helpful to stay in touch with other PSCers who have gone through the same experience. Patients were urged to seek professional psychological help if necessary. It will all be worth it in the end, they said.

Reported by Caroline Vanneste

Donna Jean's Story

Donna Jean who was unable to attend the conference and participate as a panelist for this session, sent her notes which were integrated into the panel discussion. Her informal notes painted such a vivid picture of her pre and post transplant experience that we asked her to consolidate them for The Duct. Donna Jean had her liver transplant in June 2010, a few weeks after the Hartford Conference.

I received my liver from the New England Organ Bank. I stayed eleven days in the hospital. My best advice PRE-transplant is to ALWAYS have health insurance. NEVER EVER be without coverage. My best advice POST-transplant is to maintain good nutrition to support the body's healing process. Healing post-transplant is directly proportional to the amount of protein you eat. Every medical professional who saw me at the hospital gave me the same message: It is critical to eat a minimum of 100g protein each day, that is, twice the normal daily protein requirement.

After a major abdominal surgery, hunger is non-existent, so eating was a huge challenge. In order to provide the protein the body needs to heal, it is critical to hit the protein targets every single day and keep

up this regimen during several months. It was too much food to physically eat, so I used whey protein powder and made fruit drinks in the blender. I kept a food journal and measured my food to make sure I was getting enough of the desired nutrients.

My healing milestones were the following: I first walked after two days after moving out of intensive care. Due to the thirty extra pounds resulting from fluid retention and swollen joints, walking was a challenge. When I returned home, my aunt took care of me for two weeks. She only left when I could cook, clean, bathe, walk by myself, and drive myself to my clinic appointments.

Here is what I recorded in my Tx Healing Journal:

Day 1-11: Hospitalization - lost circadian rhythm (wake/sleep cycle like an infant's, no sleep pattern)

Day 4: Discontinued pain medication (I did not need it); did not require any pain medication after leaving hospital; managed all pain with a hot water bottle - it worked wonders!

Day 12: 2 naps, 4 walks for total of 2.0 miles, ate ~140g protein; bile stopped flowing to the external drain

Day 14: Can take full care of myself: bathe, cook, do chores, exercise. All I am doing is eating, walking, drinking water, sleeping, chores. It is a full and exhausting schedule - no time for anything else

Day 18: able to walk a total of 5.0 miles in many small walks; sleep ~12 hrs + naps

Day 26: Staples removed from incision (~30)

Day 23: First drive

Day 30: Can read the Economist again (it has been hard to concentrate due to anemia); have established wake/sleep cycle

Day 45: Able to walk 7.5 miles/daily; pretty much all pain gone now; still anemic and getting tired

Day 50: First mountain climb: Mt Cardigan, NH (elevation: 3121ft; still had bile tube in; selected an easy path 'suitable for a child's first mountain hike') Photo attached

Day 69: Bile tube removed; no issue (cannot swim until tube is removed)

Day 79: First swim (Walden Pond, Concord MA) - no pain- full range of motion- just wonderful

Day 100: Allowed to begin arm exercises starting at 1lb. Walking 7mi/day

Day 115: Walking 9mi/day; continue arm exercises at 2lbs, allowed to begin abdominal exercises - can do barely 10 crunches

Day 125: Allowed to return to work part-time to 20 hrs/week (due to anemia improvements), includes 1hr+ commute. Exhausted!

Day 151: First whitewater canoe trip on easy class-2 white water. I did everything myself except lift my 50lb boat to/from the car roof

Day 160: Able to do 100 abdominal crunches, 8 lb arm exercises

Day 180: Full time at work, able to shovel snow (and lots of it!); still get tired and sleep ~10hrs/day.

Day 180 to now: Still get tired, can do most activities - fatigue unpredictable. Life is good Post-Tx!!



Medications

I was discharged from the hospital on ~10 medications. Within the first 3 months, all were tapered off except the immunosuppressants, Prograf and Cellcept, and the anti-viral for prophylaxis (discontinued at 6 months).

My Tx center tapers patients off steroids before discharging them from hospital. They also recommend taking fish oil, as that ameliorates some of the toxicity of the Prograf on the kidneys. It is key to drink lots of water to help protect kidneys.

Hospital Stay and What to Expect

I only needed my health insurance card, sports sandals with adjustable straps, warm socks, comfort items, toothbrush, soap, and comb.

Ladies - put your hair into an easy-to-care for style (braid) - you are not going to not be able to brush your hair for a few days. Don't bother packing any clothes. Socks and good footwear are the only clothing you need. You DO need chapstick. Some of the tubes can irritate your lips, but they are soon removed. The only thing in the world I wanted immediately after the surgery was chapstick.

Put it all in a "GO" bag, along with a copy of your medical records and all contact phone numbers. Make copies of these to give out as handouts. Include all your doctors, family, friends. The sports sandals helped me get up and walk - they are washable (important because the gut has a lot of issues), they provide good foot support (hard to walk with IV, bloated from fluids, huge incision making movement difficult), and have adjustable straps to allow for the huge amount of swelling.

Get a pre-paid phone or have someone manage your phone minutes. My largest post Tx bill was my Verizon wireless bill, because I went over 'minutes.'

Have a mental plan for what you want to do when you wake up from the surgery. Mine was to do an evaluation to check for myself whether I had anything really bad happen (like a stroke), so I planned to wiggle each body part to make sure I could move them, read something as soon as possible, and try some simple mental arithmetic.

Then I knew that it was important to walk as soon as possible, no matter the pain. I woke up earlier than expected, and did those things (couldn't add very well), and passed the time by giving thanks for all that was wonderful in my life.

Entertainment Options

Reading material is important to get your brain working. I could read the newspaper. It was very hard to concentrate due to all the medications. But your main job is to get better. So try to eat when allowed, walk as much as possible, socialize on the phone/visitors, and do your best to get out of the hospital. You'll have a lot of visitors from the Tx team. It is hard to get rest in the hospital, and everything takes lots of energy.

Choosing a Transplant Center

My Tx center chose me. The first Tx center I went to, a major Boston teaching hospital affiliated with my hepatologist, rejected me because my MELD score was too low. Lahey Clinic Tx Center accepted me for listing: their philosophy (different from the other center) is that the MELD score is meaningless in PSC, and that any small problem could cause a dramatic change in status. They felt listing 'early' (e.g. low MELD) was the most prudent option. One can sit on the list for years, but it is important to be on the list .

They have Tx'd ~100 folks with PSC there, more than any other hospital near Boston. They have a 'team' approach, which made me feel a bit uncomfortable, until I saw them working together. This is an amazing team, and I don't think I could have done better. I had a very positive experience, and would highly recommend shopping around if possible. It is important to remember that different places have different approaches. I felt safe with the medical team because they were realistic, aggressive, and very supportive.

Managing the Wait for "the Call"

Pack the 'GO' bag, and always carry it with you. Live with your cell phone. Get a spare battery. Keep them both charged. Plan activities within your 'radius', and try not to feel like it is a cage - the illness is the cage, and causes physical limitations that make it unlikely to get outside of the three-hour radius. Practice your response to "the Call" - perhaps do a 'dry run'. I didn't do this. My IQ dropped in half when I got 'the call' - I don't know how I managed the decision, and somehow got through it all. Expect this. Have someone else drive you to the hospital if possible. I didn't expect 'the call' because my brother was being worked up to be a live donor. But it happened (on my brother's birthday), and it was amazing that it all worked out.

Support System

Mine was not very organized. I didn't expect to get 'the call,' and didn't quite have a phone/email tree set up. I recommend organizing the phone tree/email list ahead of time. Prepare for a wide variety of reactions: Some will be very supportive, and others will distance themselves. Just like sharing your diagnosis - it always was a surprise to see how people reacted.

Educating Yourself Ahead of Time

Talk to folks that have experienced this. This was the single most helpful thing that helped manage my fears. At last year's PSC conference, I met wonderful people who shared their Tx experience with me. At that time I was researching cholangiocarcinoma and live donor transplantation. It never occurred to me that I would receive a gift from the 'List' just a few weeks after the conference. I used all the advice folks shared with me. The 'living proof' helped me know that there would be a few really bad days in the hospital (maybe 2 or 3), and that the whole thing was completely doable by a scaredy-cat, such as myself. Although folks did tell me that I would feel better after waking up from the surgery, I didn't really expect it. I had no idea how sick I actually was until I woke up from the surgery and felt a warm, deep feeling of wellness which I hadn't

We are pleased to extend an all-year discounted rate to members of PSC Partners Seeking A Cure at either **Doubletree Hotel by Hilton** – Mayo Clinic Area or the **Hilton Garden Inn** Downtown Rochester, during any of your travels to Rochester, Minnesota. Please select one of the reservation links below to begin your reservation process for the 10% discount. <http://doubletree.hilton.com/en/dt/reservations=RSTDTDT&corporateCode=0002757785>



OR <http://hiltongardeninn.hilton.com/en/gi/reservations/index.jhtml?hotel=RSTRHGI&corporateCode=N2757786>

Caregivers: Helping Us Help Others

Panel of Caregivers

Moderator: Mike Pearlman

Panelists: Joanne Grieme, Scott Malat, Karen Pearlman,
Eve Jedrzejewska, Caroline Vanneste

Panel

Mike Pearlman - Moderator and father caregiver

Joanne Grieme - Mother caregiver of adolescent/young adult three-time transplant recipient

Scott Malat - Spouse caregiver

Karen Pearlman - Sibling caregiver

Eve Jedrzejewska - Mother caregiver of pediatric daughter

Caroline Vanneste – Spouse caregiver of adult transplant recipient

This breakout session was well attended and explored the many complex issues associated with being a caregiver to a family member with PSC.

Overwhelmingly present was the theme that there exists no single path or prescribed instructions on being a caregiver. It is apparent that each situation and relationship presents a unique set of challenges and opportunities for providing meaningful support, respect and assistance for both the caregiver and the family member with PSC. It is also clear that the role of caregiver changes according to the changing needs or absence of needs of the patient.

Another recurrent theme during the session was the importance of refraining from defining the patient through PSC. Always asking the patient, “How are you feeling?” only reinforces the disease and does not help the family member having PSC. It was even pointed out that our use of the term PSCer may also contribute to this unintended consequence. It is widely known to all of us that this sometimes invisible disease invokes a wide variety of similar questions from friends, teachers and other relatives who do not understand how sick our child, spouse or friend can be. However, we must strive for a life that includes and highlights moments of normalcy.

Most caregivers experience a tremendous frustration about not being able to fix their loved one’s PSC. As caregivers, it is vital to maintain a sense of well-being and to understand that we cannot be everything to everyone all the time. In our role of caregiver, it is critical that we take care of ourselves physically and emotionally.

The following are concrete suggestions for helping our family member with PSC:

- Providing emotional support through listening
- Helping find moments of normalcy with family, within marriage and in relationships
- Focusing on the person and on what makes him/her happy versus focusing on the disease
- Not allowing PSC to control family relationships
- Asking what role our PSC family member wants us to play
- Educating ourselves about the disease, treatment plans, options, and advocacy
- Keeping surprises to a minimum, especially with children and young adults
- Preparing our patients to become their own advocate for care, diet, treatment, legal and insurance matters
- Learning to trust them with their own clinical condition assessment and allowing them to know and respond to the signals of their own body
- Considering executing power of attorneys and/or advance directives to allow caregivers, if necessary, to have the right to pay bills, deal with insurance companies, doctors, labs, etc.
- Understanding that guilt and resentment, while natural, will not foster the necessary bond for successful caregiver-patient-family relationships
- Recognizing the need for privacy

These suggestions only scratch the surface of this subject and clearly are applicable in some cases and not in others.

The bottom line is that there is no “normal” and that we must work hard to keep the lines of communication between patient and caregiver open and honest. While sometimes difficult, this kind of openness is absolutely critical. Open communication promotes the trust and mutual respect needed to maintain a positive role as caregiver.

Reported by Ken Shepherd



Medical Therapies on the Horizon

Drs. Bowlus, Selmi, Hirschfield, Melum and Zern (Dr. Mark Zern, Director Transplant Research Institute, Professor of Medicine, UC Davis)

This PowerPoint presentation is available at ["Medical Therapies on the Horizon"](#)

Dr. Christopher Bowlus – Targeting inflammatory cells

Whenever there is an injury, lymphocytes, guided by chemoattractants, leave the blood vessel to address the area of injury. Part of the process of PSC causes immune cells to be misdirected to the liver. Two molecules of interest, which home lymphocytes to the gut, are $\alpha 4\beta 7$ and CCR9. These molecules interact with MADCAM and CCL25 respectively. This process may be relevant to PSC, as it is thought that the gut lymphocytes that should pass through the liver, are misdirected into the liver instead.

Drugs that may be considered:

- *Natalizumab* is an $\alpha 4$ blocker currently in use for multiple sclerosis (MS) and is minimally effective in Crohn's disease. This drug is no longer being pursued because of the significant risk of developing progressive multifocal leukoencephalopathy (PML).
- *Vedolizumab*, an $\alpha 4\beta 7$ blocker, is showing promise in clinical trials, with much lower risk of side effects, as it specifically targets $\alpha 4\beta 7$.
- *CCX-282*, a CCR9 blocker, is in phase II study for Crohn's and is also showing promise.

With funding from PSC Partners Seeking a Cure, novel molecules are being developed as oral medications to specifically block ($\alpha 4\beta 7$).

Dr. Selmi – Probiotics and Antibiotics

Thousands of bacterial and fungal species generate a complex environment that grows and co-evolves with the host. These participate in digestion of nutrients, protection of mucosa, and development of a healthy gut, and evolve into a balanced mucosal immune system. Probiotics are "live microorganisms which, when consumed in adequate amounts as part of food, confer a health benefit on the host."

Probiotics produce antibacterial substances, secrete mucosal cytoprotective agents (protects cells from noxious chemicals), inhibit pathogens, enhance barriers and immune roles of intestinal epithelial cells, and regulate mucosal immune responses. They enhance host innate immunity, increase anti-inflammatory cytokines (cell-signaling protein molecules), suppress pro-inflammatory cytokines, and up-regulate host defenses against infection.

There is evidence from trials using VSL#3, that probiotics could improve IBD disease activity. This could help PSC patients, as 90 percent are affected by IBD. Currently, there are no clinical trials using probiotics in PSC patients.

Vancomycin is a bactericidal antibiotic poorly adsorbed by the intestinal tract. Given orally, it acts on

PSC + IBD pediatric patients treated only with sulfasalazine and vancomycin reported promising data. When the patient was re-challenged (taken off, then put back on Vancomycin), there was rebound in GGT & ALT. Due to the small number of patients involved in this clinical trial, the evidence is far from conclusive. However Vancomycin may prove to be useful in certain cases.

Dr. Melum – Novel drug targets from genomic studies - anticipating the future

As stated in previous presentations, there may be multiple versions of PSC, and along with multiple environmental triggers, multiple drugs may be required. We need to group the different types of PSC and identify individual treatment.

Due to the relatively rare nature of PSC, a good strategy is to “hitchhike” on related, more prevalent disease studies that may have applicability to PSC.

Genotyping chips can be used to categorize the various types of a disease. An example is the ImmunoChip Project, where the genetic information and conditions (multiple sclerosis, asthma, IBD, rheumatoid arthritis, and others) of 4000 PSC patients are being used to determine whether a medication for one of the related and far more common conditions may prove to be useful for PSC. For example, Ustekinumab (anti-p40) has shown to be effective for psoriasis, suggesting that it might also work for Crohn’s Disease and possibly for sarcoidosis. These conditions often occur with PBC, suggesting that anti-p40 would be worth trying for PBC. Hopefully, we can find similar situations for PSC.

Dr. Hirschfield – Urso (UDCA) and beyond

It is to be noted that it doesn’t have to take long from discovery of a drug application for a disease to its use as treatment, especially if the drug has been licensed for other uses. Biotech companies are very interested in expanding the use of an existing, licensed drug, as the safety data has already been developed.

Bile acids are not just detergents to solubilize fats in the intestine, but are also signaling molecules. Some of the damage caused during cholestasis results from retention of toxic bile acids, and not just from lack of flow. Consequently, a treatment can comprise ways to channel out toxic bile acids. Obeticholic Acid (or INT-747) is similar to Urso and is a very strong FXR agonist. In animal models, it is anti-cholestatic. When INT-747 is infused, bile flow increases.

In a placebo-controlled clinical trial, INT-747 was used on stable PBC patients not on Urso. Patients were not on Urso either because they had not been treated with Urso prior to the trial or because Urso had not been effective for them. For PBC, the study showed INT-747 to be as effective as Urso in reducing alkaline phosphatase. A rapid drop in GGT was observed (low GGT = less toxic bile). A major side effect was pruritus (itching), but it was dose dependent, and lower doses turned out to be just as effective.

In order to improve the time to release for new medications, an “add-on” license is sometimes pursued. For example, adding Obeticholic acid to Urso, an approved PBC medication, could shorten the wait.

This is another example of how PSC research could hitchhike on PBC, a more common disease than PSC. This clinical trial should hopefully move to phase III this year.

Dr. Zern – Stem cell transplantation

Liver cells could be directly injected into the portal vein, colonizing the existing liver. Alternatively, much like kidney dialysis, a bioartificial liver external to the body could be developed. In this process, embryonic stem cells are “tricked” into becoming liver cells. Currently 80-90 percent of the stem cells develop into liver cells. Just like hepatocytes (liver cells), these stem cells express albumin.

Stem cell transplantation is being researched in Egypt. Stem cells are derived from adult bone marrow, then activated into liver cells and re-infused into the patient. No immunosuppression is required since the new cells are generated from their own bone marrow. Patients in end stage liver disease, primarily hepatitis C, showed improvement. Although this process is not a substitute for whole organ transplantation, it may considerably extend time to transplant.

Reported by Arne Myrabo

The Effects of Diet on Inflammation

Charles Stephensen, PhD, Adjunct Professor, Department of Nutrition, UC Davis,
USDA ARS Western Human Nutrition Research Center Scientist

Dr. Stephensen's PowerPoint presentation is available at "[The Effects of Diet on Inflammation](#)"

Dr. Stephensen began with a brief description of the programs at the Center, which include human intervention trials, experimental diets, and effects of diet on physiological function, bone density, and inflammation. He followed with a definition and description of inflammation and its stages. The majority of the presentation focused on specific dietary components and their effects on inflammation.

Pre-biotics are non-digestible food components that are thought to promote beneficial bacteria.

Probiotics are actual organisms (good bacteria) that are ingested in supplements or in foods such as yogurt. One summary of existing studies indicated that probiotics may decrease the duration and severity of gastroenteritis by up to 50%. Probiotics may also reduce the initial damage from GI infections and improve the resolution of inflammation.

Vitamin D (often called more specifically D3) seems to improve the ability of macrophages to kill bacteria. Vitamin D is also involved in the resolution of inflammation. The speaker stated that some individuals need no vitamin D supplement if they get good sun exposure. For most people, 600 IU is a typically recommended daily base level, but some physicians may recommend up to 2,000 IU daily.

The speaker showed a diagram of the essential chemical difference between omega-3 and omega-6 fatty acids. Omega-3 fatty acids appear to help resolve chronic inflammation, for example in coronary arteries. Omega-6 fatty acids are believed to promote inflammation.

There is some evidence that mild exercise improves inflammatory resolution. Exercise stimulates the production of cortisol, which is important in inflammation resolution. High levels of cortisol are anti-inflammatory.

Q & A:

Is grass-fed beef higher in omega-3 fatty acids than corn-fed beef? There may be some research indicating that, but the key is to maintain a balance, which you can do by eating a mixture of foods.

Does the speaker have any advice on nutrition controversies raised by books such as *The China Study* or *Good Calorie, Bad Calorie*? He was not familiar enough with those books to comment specifically – his nutrition advice is already incorporated into his presentation.

Is Vitamin D2 or Vitamin D3 better to take as a supplement? He prefers D3 because it is absorbed better and lasts longer in your system.

How reliable and safe are fish oil supplements? He thinks they are generally safe.

How good is the manufacturing quality of probiotics? He was unaware of large problems.

Reported by Stephen Hatchett

Financial and Insurance Considerations for Transplant

Christina Allman, Financial Coordinator Supervisor, Transplant Program,
University of California Davis

Ms. Allman's PowerPoint presentation is available at ["Financial and Insurance Considerations for Transplant"](#)

- Financial Coordinators
 - They are responsible to make sure you are financially prepared.
 - They will review insurance coverage with you.
 - Keep the phone number of the Financial Coordinator in your cell phone.
 - Your financial coordinator is one of your most important contacts.
 - They will prepare you for the financial component of your transplant by
 - Providing education
 - Assisting with determining strategy
 - Making long term plans
 - Investigating insurance

- There are 500 coordinators across the country.
- At UC Davis the transplant patient is assigned a financial coordinator to assist the patient long-term.
- They will serve as another set of eyes to assist the patient.
- You need to review your insurance coverage prior to listing for transplant, to find exclusions.
- In a transplant situation, your insurance coverage is considered a life long commitment.
 - What type of retirement insurance coverage do you have?
 - How long will your employer pay your insurance premiums?
 - What type of short and long term disability insurance do you have?
- Every insurance company has a transplant team.
 - Someone you can go to directly to get questions answered.
 - Travel and lodging should be able to be reimbursed.
- Medicare funding
 - Getting disabled under Medicare for liver diseases is difficult.
 - There isn't much knowledge in the Medicare field on liver disease.
 - Medicare payments don't start until age 65 or if you become disabled.
 - You cannot begin receiving disability income until 2 years after Social Security classifies you as disabled.
 - Begin the process to become classified as disabled as soon as possible.
 - If denied, appeal! You may have to appeal 2-3 times.
 - Have an extensive list and information about your disease.
 - Make sure that you have a letter from your doctor that is treating you when you go to apply for disability.
 - Any letters that you can get from the physicians that are treating you will help you.
 - List every single doctor that has treated you for your disease.
 - Doctors, nurses and financial coordinators can help you formulate your petition for disability.
 - It may take a team to assist you to get disability.

Reported by Bruce Blum

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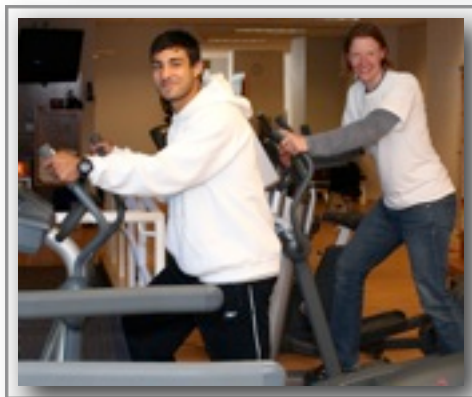
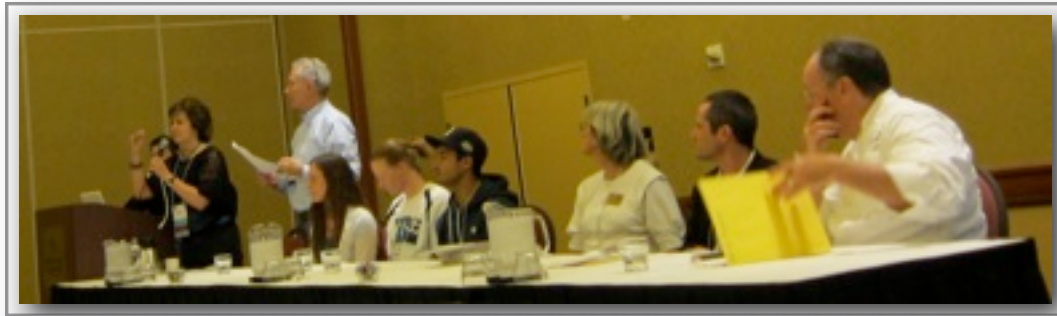
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Pre-Conference Friday Afternoon Break-Out Sessions

Healthy Living Perspectives:

Nutrition, Exercise and Alternative Therapies



Integrating Chinese Medicine & PSC: Accessing the Best of East & West

Misha Ruth Cohen, Doctor of Oriental Medicine, Research Specialist for Integrative
Medicine at the Institute for Health and Aging
University of California San Francisco

Dr. Cohen's PowerPoint presentation is available at [Integrated Chinese Medicine and PSC](#):

Dr. Misha Ruth Cohen was the keynote speaker for the Friday afternoon program, *Healthy Living Perspectives: Nutrition, Exercise and Alternative Therapies*. She shared insights from her 34 years of experience in Asian Medicine.

She started her presentation by asking the audience of PSCers and caregivers whether they used any form of complementary and alternative medicine along with traditional western medicine. About 75 percent of the audience did. About 50 percent used a form of Chinese medicine, that is, acupuncture, Chinese

relaxation, and/or Chinese exercise.

She explained that integrating Chinese and Western medicine had the advantage of combining scientific evidence coming from western medicine and thousands of years of study and practice.

She compared the basis for both systems of medicine and explained that the goal of Chinese traditional medicine was to help restore the body to balance by working on energy flow level to affect mind-body-spirit. Chinese medicine heals and prevents disease by correcting imbalances that signal illness and pain.

Dr. Cohen warned the audience against indiscriminate and uninformed use of Chinese herbs and against the dangerous practice of buying herbs off the health-store shelf. No different from any other medication, some herbs have negative interactions with western medications, and some Chinese-herb combinations have unwanted interactions. She stressed the importance of choosing practitioners qualified in both Western and Chinese medicine *and* very importantly, in liver disease.

When used in conjunction with Western medicine and by expert practitioners, Dr. Cohen said that Chinese medicine could offer complementary ways of dealing with PSC. She explained how Chinese medicine, practiced and evolving over thousands of years, is a complete medical system with its own diagnostic methods, modalities (a pattern of symptoms that is used for diagnosis), prognoses and treatments.

She gave the audience a brief overview of the premises underlying Chinese medicine. The basis of Chinese medicine lies in the way it views the body as an energetic system in dynamic balance. “*Qi* (chi)– translated as energy or life force,” she said, “flows in a regular pattern through a system of channels, or meridians, through the whole body. When the flow of *Qi* is unimpeded, there is harmony, balance, and good health. When there are *Qi* blockages or too much or too little *Qi*, there is imbalance, which can lead to disharmony and disease.”

In Chinese medicine, mind and body are not considered as separate entities. It is all about maintaining a harmonious flow of *Qi*. The liver allows *Qi* to move energy. In the liver, *Xue* (chuy, translated as *blood*) is stored, and if the flow of *Xue* is blocked, the vital energy *Qi* cannot flow properly. This flow controls aspects such as emotions, eyes, skin, hair, tendons and digestion.

In traditional Chinese medicine, diagnosis is made by looking at the shape and size of the tongue, the pulse, (and this is no simple matter, as practitioners consider 28 qualities of the pulse for diagnosis!), color of complexion, gait, emotional state, and more.

In her practice, Dr. Cohen incorporates acupuncture, nutrition consults, meditation, exercise and Chinese herbs that are carefully tested for bacteria and heavy metals. She constantly re-assesses her treatment recommendations when therapy changes are prescribed by Western medicine. At all times, she works together with the patient’s Western practitioners.

Dr. Cohen said that acupuncture, the ancient art of inserting sterilized needles into specific points in the body, has been shown to stimulate endorphins and affect serotonin levels. It is used for pain relief and for its anti-inflammatory properties. She remarked that acupuncture could decrease fatigue, upper right

quadrant pain, nausea and pruritus. She said that acupuncture might also lower elevated transaminases.

In her clinic, dietary therapy focuses on increasing energy and balancing body *Qi* and *Xue*. Massage includes meridian pressure such as *Shiatsu*, *Tui Na* or muscle massage. She also uses *Moxibustion*, which is the burning of the common fragrant herb *moxa* (mugwort) over areas of the body for stimulation and to relieve pain. Her therapy includes martial arts or gentler movement such as *Tai Chi*, *Qi Gong*, and *Yoga*, or gym workouts and aerobic exercise.

When asked whether Chinese herbs had been tested with clinical trials, Dr. Cohen explained that it would be difficult to perform clinical trials for the thousands of Chinese herbs that are used singly or in specific and varying combinations for specific conditions and for varying levels of disease.

She stressed the importance of taking control over one's health and pursuing informed self-care that would include keeping a daily journal, following dietary guidelines, performing self-massage and moxibustion, meditating and taking the right nutritional supplements. It was informative to have the perspective of an experienced and professional Chinese medicine practitioner on complementary treatments for PSC.

Dr. Cohen said she would welcome interested PSCers to contact her at 415-861-1101, ChinMedSF@aol.com

Reported by Rachel Gomel



HOME HEALTH

Cindy Campbell RN, BSN Fazzi Associates, Inc.

Cindy Campbell's PowerPoint presentation is available at <http://pscpartners.org/PSCConf11/PDFs/Campbell-PSC%20Partners%202011.pdf>

Cindy Campbell works on creating and spreading home care policies nationally. Her approach to home care is down-to-earth and is based on the premise that healthcare happens naturally in the home every day. When outside help is needed, however, we often don't know our options.

In fact, what we should know is that the hospital can come home if the need arises. Diagnostic procedures, treatments requiring high skill, occupational and physical therapy are all within reach within the home. To receive these services, the patient must have "homebound" status. The kind of care Cindy talks about is Medicare Certified and includes monitoring and teaching interventions and involves skilled nurses, physical and occupational therapists, social workers, and home health aides who all work as a team in accordance to the plan of care established by the physician.

Financially, the patient is covered 100 percent by Medicare. With insurance companies, these services are policy-specific and can take the shape of co-payment. In the case of Medicaid which is state-specific, most professional care is covered as long as it is pre-authorized.

It is important to note that skilled disciplines and medical supplies are covered while durable medical equipment (DME), and personal “custodial care” are not.

Hospice and palliative care at home includes nursing, social services, spiritual support, and medical and personal care. These special care providers all work collaboratively. In this case, the patient is covered 100 percent by Medicaid. With insurance companies, coverage is policy specific. Hospice benefit pays for medication, durable medical equipment (DME), and intermittently, for skilled disciplines and personal care.

Cindy suggested a home health checklist for PSCers. She advised PSCers to identify milestones of disease status, coordinate lab screening and testing as indicated by best practice guidelines, list all medications/supplements and update those as needed, and look for symptoms that may be causing flares. By finding patterns, it may be possible to interrupt problems.

Cindy said that chronic disease management evolves directly with technology, and named PSC Partners as powerful evidence of this phenomenon. Technology has created an education and support system that knows no geographical boundary and that abolishes distance. The PSC Partners group is the consequence of technology and is able to help its members create the very best environment of caring.

Cindy emphasized the importance of self-care through making good nutrition choices, taking required medication and supplements, and making sure to include exercise, rest, intellectual stimulation, and activities that are a source of joy to the PSCer. Ultimately, we must each be our own best friend by knowing how to advocate for ourselves within the health system by keeping documents up to date, by seeking competence, and by being vigilant about our insurance status. We must always strongly and carefully advocate for ourselves.

Reported by Rachel Gomel

**Set up Your Own Fundraiser for Save the Day,
October 6-8, 2011!**

Get the gang together! Here's how to help find that cure for PSC by funding research.

There are loads of great suggestions for local fundraising activities at our website:

<http://pscpartners.org/fundraisers>.

Register your idea with Sandi Pearlman at fundraising@pscpartners.org.

Following Dr. Cohen's presentation, attendees chose two breakout sessions from the following: *Kinetics for PSCers*; *Connecting Food, Health and the Environment*; *The Glories of Eating Seasonally and Locally*; *Gardening - Good for the Gut, Good for the Soul*; *Yoga: Feeling is Believing*; *A Gluten Free Update*; *Coping with a Gluten Free Diet in Real Life*; and *An Integrated Chinese Medicine Case Study in PSC*.

Julianne Vasichek and Cameron Wohl's PowerPoint presentation, *Kinetics for PSCers*, is available at <http://pscpartners.org/PSCConf11/PDFs/Kinetics.pdf>

An Overview of Disability Law

Becky Long, Esq., PSC Partners Board Member and Legal Counsel

The answer to whether a patient is entitled to disability protection is a complicated one, as it varies according to different laws. It depends on the tasks that one is able to perform and not on diagnosis. Below is a brief overview of federal statutes. For individual questions, please visit www.advocacyforpatients.org. Jennifer Jaff of *Advocacy for Patients with Chronic Illness* provides free information, advice, and legal advocacy services in many areas of disability law.

Family Medical Leave Act (FMLA)

For those who need time off from work to take care of themselves or of an immediate family member, the Family Medical Leave Act provides twelve weeks of unpaid leave every twelve months. This requires proof of a "serious health condition" by certification from a physician. This leave can be taken intermittently, but it may run concurrently with vacation/sick time/short-term disability. FMLA applies only to employees working for a company of fifty or more employees and is limited to employees who have been employed in the organization for more than twelve months. The request for FMLA leave requires a thirty day notice or must be made "as soon as practicable." The employer MUST maintain group health insurance for an employee during FMLA leave, though payment of premiums may be required by the employee.

When should one involve HR? This is a judgment call, but it is advised to be proactive when possible. One must think of FMLA as job protection/security. It is important to keep an HR file including copies of all correspondence and forms with their specific dates. Keep in mind that most managers do not act out of malice but are simply uninformed.

Americans with Disabilities Act (ADA)

Coverage under the ADA requires 1. *a physical or mental impairment* 2. *that substantially limits one or more major life activities*. Sleep, digestion, bowel, and immune functions are some of the functions that define a major life activity. *Qualified Individuals* are those able to 3. perform the "essential functions" of their job 4. with or without "reasonable accommodation." 5. A "Reasonable Accommodation" must not create an "Undue Hardship" on the employer.

The undue hardship determination requires a case-by-case analysis of the patient's condition, job duties, and the employer's ability to make an accommodation based on the size and financial status of the company. If a request for a specific accommodation is made, there is a chance that some other accommodation will be offered instead. One must negotiate, as this is an "interactive process."

Long-Term Disability (LTD) It is almost impossible to find private Long Term Disability Insurance outside of an employer-based group plan for a chronic illness. If offered by the employer, it is advisable to opt for this coverage. It is important to note that most private LTD requires application for SSDI.

Social Security Disability (SSDI)

Most people do not want to be on disability insurance. It is a tough decision to admit inability to work. With a salary of less than than ~\$1,000 a month, it is possible to work and still receive SSDI. On the other hand, it will be difficult to convince SSA of disability while working. Please consult www.ssa.gov and read every line carefully! To be eligible, sufficient payment must have been made to Social Security during active work years. Dependent children may also be eligible.

The following is the five-part test for SSDI: 1) Is the claimant currently employed? 2) Does the claimant have a severe impairment, that meets a listing for automatic eligibility? (Currently, PSC is not an automatic listing.) 3) Does the claimant have a medically equivalent impairment? 4) Does impairment prevent the claimant from doing his or her job? 5) Does the claimant have "residual functional capacity" to perform any job?

At the *Application* stage, 70 percent are denied on first try. At the *Reconsideration* stage, another round of rejections can be expected. Then follows a *Hearing* before an Administrative Law Judge. At that time, it is advised to retain an attorney. (Please consult the National Organization of Social Security Claimants at www.nosscr.org). The next phases include *Appeals Council* followed by the *Court System* (often remanded back to ALJ).

When should one apply for SSDI? One should apply as soon as s/he is unable to work. The application process can take, on average, two years if a hearing is required. One can expect to be without an income during this time. Benefit checks begin six months after the onset of disability, and retroactive payments are given if the approval process is lengthy. Medicare begins once SSDI benefits have been received for two years. It is important not to be discouraged! About 50 percent of those who appeal are granted benefits.

Social Security Income (SSI)

For those who are too young or not eligible for SSDI, it is advised that SSI be considered. The disability determination is the same as for SSDI. Benefits begin the first month after approval. It is also possible to be considered "presumptively disabled" and receive benefits during six months while the application is being processed. In most states, SSI comes with immediate eligibility for Medicare.

Educational Equity

Under the *Individuals with Disabilities in Education Act* (IDEA), all children are entitled to a free appropriate public education. If by reason of "health impairment," a child needs "special

education and related services,” he or she is entitled to an Individual Education Plan. Under Section 504, disabled students cannot be denied the benefits of any program receiving federal financial assistance and must be granted “reasonable accommodations” (same standard as the ADA). Students are covered by the ADA in post-secondary and private schools.

Appealing a Denial of a Health Insurance Claim

The insurance company’s denial letter MUST include 1. a specific reason for the denial 2. a specific reference to pertinent plan provisions and 3. the steps needed for submitting a claim for review. The applicant MUST be given access to his or her files and be provided copies. It is important to be one’s own advocate! Persistence pays off, as 70 percent of health insurance claim appeals are successful. It is important to write a letter from the applicant’s perspective and to make sure that the physician’s letter include objective medical evidence, medical records, and articles and journals (www.psc-literature.org). Take appeals seriously and exhaust all internal appeals. This is the best chance for success because the standard of review is higher in court.

Life Insurance

Most PSCers are denied private policies. Be sure to designate a beneficiary if you have a policy available through your employer. Get a policy for your spouse if possible. Tell loved ones to consider purchasing life insurance while healthy. For questions, you can write to Tim Reid of Northwestern Mutual at trreid365@gmail.com.

When to consider getting an Attorney?

1. For SSDI appeal, a professional presentation at the hearing stage is needed to cross-examine medical and vocational experts. 2. For the appeal of an LTD Claim an attorney is advised because of the amount of money that is at stake. 3. For the External Appeal of a Health Insurance Claim, it is advisable to negotiate a contingency rate and seek an attorney willing to handle the appeal only, as opposed to an attorney who wants to litigate.

Becky Long

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January 1 to May 31 2011

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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.

For summaries of
the general
sessions, please
read Part I of the
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PSC Partners Seeking a Cure is a 501(c)3 nonprofit foundation that endeavors to find a cure for Primary Sclerosing Cholangitis.

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

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.

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