

# PBC connections

A Canadian PBC Society Newsletter

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## ADVANCES IN IDENTIFYING THE GENETIC BASIS OF PRIMARY BILIARY CIRRHOSIS ■ *Dr. Gideon Hirschfield, Dr. Jenny Heathcote and Dr. Kathy Siminovitch*

Over the past years, progress in identifying the genetic basis of Primary Biliary Cirrhosis (PBC) has been – in a word – spectacular. Thanks to the generosity of the PBC Society and its patient community, the efforts of hepatologists across this country, a strong network of international collaborators and an excellent research support team, much of this progress in PBC genetics originates from the Canadian research community. Our research team was, for example, the very first to carry out a broad (genome-wide) screen for genes conferring risk for PBC. By this means, three of the key genes that predispose to PBC were identified, findings published in the *New England Journal of Medicine* in 2009. Via close collaborations with the Mayo Clinic and University of California at Davis in the United States, our team went on to identify four more of the genes involved in risk for PBC. In our most recent studies, we have capitalized on new genetic technology and our growing cohort of PBC patients (one of the largest worldwide) to identify another important gene for PBC and to fine-tune our knowledge of many other PBC susceptibility genes. Thus in the past three years, we have witnessed and helped lead an unprecedented increase in knowledge of the genes that predispose to PBC.

Our research is, of course, far from finished. We now need to define the complex series of protein and cell interactions whereby PBC risk genes alter an individual's immune and other biological systems so as to cause disease. We must also connect our new genetic understanding of PBC to understanding of why some PBC patients respond to certain therapies and some do not and why the disease progresses more rapidly in some individuals than in others. Such knowledge will allow our team and others to develop more effective treatments and new strategies to mitigate risk and improve outcome for all those predisposed to or affected by PBC.

We thank the more than 1000 individuals involved in our study for the participation and support that has fueled our research and created an exceptional opportunity to translate the remarkable advances in genetic technologies to meaningful impact on the health of PBC patients.



DR. GIDEON HIRSCHFELD



DR. JENNY HEATHCOTE



DR. KATHY SIMINOVITCH



# UPDATES

## “PILOT STUDY OF HIGHLY ACTIVE ANTI-RETROVIRAL THERAPY FOR PATIENTS WITH PRIMARY BILIARY CIRRHOSIS”

*Andrew L. Mason MBBS MRCPI - Professor, Department of Medicine, University of Alberta*



We are starting up our antiviral treatment study funded by Canadian Institutes for Health Research: “Pilot study of highly active anti-retroviral therapy for patients with primary biliary cirrhosis”. The study sites will be in Vancouver, Edmonton, Calgary and Halifax in Canada, as well as St. Louis University, Mayo Clinic Rochester MN, and University of Texas, South Western in the USA. The rationale for this study is based on ongoing research in my laboratory. We found evidence of retrovirus infection in patients with primary biliary cirrhosis and reported the discovery in 2003.

Since then, we found that most patients with PBC have evidence of viral infection using the gold standard technique of showing viral integration into the human genome. We have also conducted preliminary clinical studies using anti-viral therapy. We found that PBC patients treated with combination anti-retrovirus therapy experienced significant reversal of the disease process. This provides some circumstantial evidence for a retroviral infection. Although we could show significant improvements, the changes were not substantial and we are now looking for better antiviral regimens. Now we have found a mouse model with a similar virus infection that develops a similar biliary disease. Importantly, we found that antiviral therapy blocks the development of the disease in this mouse. We have used this model to find safer and more effective antiviral treatments for patients with PBC. We have now found out that a combination of highly active antiretroviral therapy with Truvada and Kaletra stops disease in the mouse and plan to use this combination to see if it works in patients with PBC. Of interest, a recent case report shows that this combination is associated with normalizing liver tests in a patient with PBC and retrovirus infection.

## INTERCEPT PHARMACEUTICALS IS ENROLLING ITS PHASE III STUDY OF OBETICHOLIC ACID IN PBC

A Phase III clinical trial in PBC, sponsored by Intercept Pharmaceuticals, Inc., is currently enrolling patients at several US, European and Canadian clinical trial centers. Over 60 centers in 13 countries are participating worldwide. In Canada, the study is being conducted at the University of Toronto by Dr. Hemant Shaw and at Hôpital Saint-Luc/CHUM in Montreal by Dr. Catherine Vincent ( e-mail: [catherine.vincent@umontreal.ca](mailto:catherine.vincent@umontreal.ca)). Enrollment in this study is progressing well and is expected to complete by the end of December 2012. Additional information about the study is available at the websites below.

To be eligible for the study, patients must be on a stable dose of UDCA (or unable to tolerate UDCA), must have an elevated alkaline phosphatase (AP) level of more than 1.67 times the upper limit of normal and a bilirubin level of less than 2 times the upper limit of normal. In this study, patients will receive OCA or placebo (a ‘sugar pill’) for 12 months, then may enter a long-term study extension during which they will take OCA (free of charge) for up to 5 years. Patients who previously participated in a trial of OCA are not eligible for enrollment in this study.

Anyone interested in learning more about this trial in Canada should contact the following individuals for further information:

**University of Toronto:** Catalina Coltescu – 416-603-5832

**Hopital Saint-Luc:** Christina Benjamin – [tinabenjamin2a@gmail.com](mailto:tinabenjamin2a@gmail.com) or 514-890-8000, ext. 35283

Also, a full list of PBC doctors participating in this study and contact information has been posted at:

**Intercept’s websites:** [www.pbc-study.com](http://www.pbc-study.com) and [www.interceptpharma.com](http://www.interceptpharma.com)

**ClinicalTrials.gov** (a registry of federally and privately supported clinical studies conducted in the United States and around the world):

[clinicaltrials.gov/ct2/show/NCT01473524?intr=%22INT-747%22&rank=4](http://clinicaltrials.gov/ct2/show/NCT01473524?intr=%22INT-747%22&rank=4)

# fund raising

## DAY AT THE RACES

The annual Day at the Races on June 10 at Woodbine Racetrack in Toronto posted another outstanding success. The event raised almost \$7,500, surpassing last year's record amount.

On an extremely hot and humid day, members, their families and friends enjoyed socializing over good food from the buffet and exciting racing. They placed bets on the horses and bids on the 27 items in the silent auction. There were also 10 items for children (or their indulgent relatives) to bid on.

Dr. Jenny Heathcote, Chair of the Francis Family Liver Clinic at Toronto Western Hospital, which is one of the leading centres of research into liver disease in the world, took us briefly through the work they are doing and updated us on her replacement following her coming retirement in 2013.

As always, thanks are due to everyone who attended and helped make the day so successful, but particularly to Schonberg Farms for their support and sponsorship of the PBC Society race, to all those who donated items for the silent auction, to Dr. Heathcote, to the ever professional and helpful Woodbine staff, and to Barbara Badstober, our hardworking President and driving force in this event.

## NEXT YEAR'S EVENT

**Sunday, June 9, 2013**

**in the spectacular Northern Dancer Room!**



## ALBERTA NORTH BARBEQUE AND SILENT AUCTION

The Alberta North Region of the Canadian PBC Society held its 5th Annual BBQ and Silent Auction Fundraiser on Saturday, May 26 at the home of Shauna and Greg Vander Well. The event was well attended and brought in over \$500 in silent auction sales and donations. The funds have been forwarded to National Office to support further research into PBC. The weather was gorgeous (if sometimes too cool for Shauna). Our guests of honour were Dr. Andrew Mason and his lovely, supportive wife, Gina. Dr. Mason told us about his latest drug study, which began the following Monday, and answered our many and various questions about PBC and treatment.

Our group certainly appreciates Dr. Mason's ongoing support. The biggest ticket item in the silent auction was donated by Gina Mason; it was her giclee print entitled "Sunrise". Thank you, Gina

For information, please contact Shauna Vander Well: [albertaN@pbc-society.ca](mailto:albertaN@pbc-society.ca) or 780-962-6217.

# UPCOMING EVENTS

## ALBERTA NORTH REGION

### The Importance of Weight & Exercise in the Management of Primary Biliary Cirrhosis

**Guest Speaker:** Dr. Arya Sharma, Professor of Medicine & Chair in Obesity Research and Management at the University of Alberta, Edmonton and Clinical Co-Chair of the Alberta Health Services Obesity Program.

**Date:** Friday, November 30th, 2012 at 7:00 pm

**Location:** The Oborowski Degner Seminar Hall, University of Alberta ADI (1-041)

**Suggested donation:** \$5 or more to cover costs. Any excess funds will be forwarded to The Canadian PBC Society as a donation toward Reaching for the Cure. Space is limited.

Register with Shauna Vander Well by e-mail [vwell@shaw.ca](mailto:vwell@shaw.ca), text 780-238-2892 or phone 780-962-6217, giving the following information: subject "PBC Guest Speaker"; your name; number attending; contact phone number. You will receive confirmation of your registration.



## ATLANTIC GROUP

**Date:** Our Fall get-together will be on Wednesday, October 17th at 11:00 am

**Location:** We have decided to try a Bedford location, the Sunnyside Restaurant, 1552 Bedford Highway, the restaurant which was formerly called Smitty's. When you enter the building, turn to your right. The following link gives the location, reviews, etc. [www.yelp.ca/biz/sunnyside-restaurant-bedford](http://www.yelp.ca/biz/sunnyside-restaurant-bedford)

If you have any questions, please let Ellen know. A response re your attendance by the week prior to the event is requested and appreciated. See you there!!

**Contact:** Ellen Hearn - [ellmac11@hotmail.com](mailto:ellmac11@hotmail.com)



# regional connections

## ALBERTA NORTH

See Fundraising section for report on the annual barbecue in May. Our next luncheon will be scheduled for the early fall; the date is to be announced.

For more information, please contact Shauna Vander Well: [albertaN@pbc-society.ca](mailto:albertaN@pbc-society.ca) or 780-962-6217.

## ATLANTIC CANADA

On Wednesday, May 23, 2012, six ladies from our group got together for a fun day of Bingo Bowling. There was spirited competition for the gutter ball prize, but Catherine Dingle won it, hands down. We then had lunch together in the food court of the Halifax Shopping Centre. A very enjoyable day!

We hope that more will join us for our lunch meeting to be held in the Fall.

For more information, please contact Judi Pemberton: [atlantic@pbc-society.ca](mailto:atlantic@pbc-society.ca) or 902-798-5554.

## BRITISH COLUMBIA

There was a surprisingly small turnout for the semi-annual Greater Vancouver meeting in June at the New Westminster Inn at the Quay luncheon. Those who did show up enjoyed good food and catching up. We discussed the possibility of going to an annual vs. semi-annual lunch in the future but will decide after seeing the turnout for the next get-together in the fall. If there are any new members or if you have any questions, please feel free to contact me.

For more information, please contact Kathryn Swift: [bc@pbc-society.ca](mailto:bc@pbc-society.ca)

## MANITOBA

On June 9, 13 of us met for the evening meal at Montana's restaurant. Some of us must like "Country" or else just because the food is good. I am a classical music lover myself.



It was so good to have a larger group which included several husbands and the daughter and granddaughter of one of our members. A welcome addition was Ruth Magnuson, the Regional Manager of the Canadian Liver Foundation, Manitoba Branch. She is willing to help us in any way possible, including offering us the use of her offices for meetings. For some newer members, the ability to share with other PBC'ers was a very important part of the evening.

As the autumn approaches, most of us will be freer to explore new avenues of possibility for learning about and understanding PBC and its impact on our lives.

I, myself, have had a very busy summer mostly with grandchildren: a graduation, an engagement party, a wedding, family visits and a major move for son #3 and his wife. Oh yes, and my larger community garden.

Trusting that all of you have enjoyed the warmer weather and vacation time.

Blessings, *Carol Seburn, Manitoba Regional Co-Ordinator*

For more information, please contact Carol Seburn: [manitoba@pbc-society.ca](mailto:manitoba@pbc-society.ca) or 204-254-5226

## OTTAWA

Our new group met on June 4. Three members participated; a fourth was expected but didn't show. We had a good chat to get to know each other and shared experiences. All were comfortable with the meet and greet approach, i.e., nothing formal, just a

friendly visit. Before we parted I proposed to meet again in the fall and the others were in favour. Maybe we'll be able to get some others out at that time, too.

For more information, please contact France Foucault: [ffoucault@hotmail.com](mailto:ffoucault@hotmail.com)

## QUEBEC

A l'annonce de la CBP en 2000, j'étais étonnée de ne pouvoir trouver un groupe de soutien francophone dans notre région. Il en fallait un, il existe maintenant depuis 5 ans et regroupe une vingtaine de personnes au Québec et en Ontario. Des liens inestimables se sont tissés au fil des ans. Je travaille dans le domaine des finances. J'aime beaucoup passer du temps en famille, voyager, jouer au golf, marcher, lire, partager un bon repas entre amis et m'impliquer socialement. Être utile donne un grand sens à ma vie simple et bien remplie. Je vous présente les membres du groupe présents au dîner du 25 mai 2012 au restaurant L'Incrédule de Longueuil. De gauche à droite : Francine A., Jill, Lorraine, Gilles, Shirley, Francine L., Brigitte, Sally, Yves, Lisette, Judith.

Francine Lamontagne, Représentante Québec, Société canadienne de la CBP



I was diagnosed with PBC in 2000 and surprised to find no French support group in our region. We needed one and it has been in existence for 5 years now, the members are from Quebec and Ontario. Invaluable links have been developed over the years. I work in the field of finance. I love spending time with my family, travelling, playing golf, walking, reading, sharing a good meal with friends and getting involved socially. Being useful means a lot to me.

Let me introduce the group members present at the dinner of May 25, 2012 at L'Incrédule Restaurant in downtown Longueuil. From left to right: Francine A., Jill, Lorraine, Gilles, Shirley, Francine L., Brigitte, Sally, Yves, Lisette, Judith.

Francine Lamontagne, Quebec Representative, Canadian PBC Society

For more information, please contact Francine Lamontagne: [QuebecRep@pbc-society.ca](mailto:QuebecRep@pbc-society.ca)

## SOUTHWESTERN ONTARIO

Hello Fellow PBC'ers! My name is Betty Van Luven, and I am one of the founding members of the newly formed "Southwestern Ontario" Support Group of the PBC Society of Canada. Currently, we are three members strong, consisting of Susan Webb, Maureen Gill and myself. We will meet every couple of months in and around the London, Stratford, Kitchener, Waterloo, Guelph areas, and are a free floating group, meaning we are very flexible to location. We also stay in touch regularly by phone and e-mail.



Left to right: Susan, Maureen and Betty

Our Vision is to support each other and have fun doing it! Let us spread the sunshine around! Our goal is to grow the Southwestern Ontario PBC Support Group.

Our group met on July 10th at an Italian restaurant in Stratford, Ontario and had a delightful meal. We shared information, laughter and friendship. It was very reassuring to finally meeting someone else with PBC and to know they understand. Our first smiles spoke lots. Did I mention we were very full? Not to mention the chocolate factory we visited afterward! :o) A great day was had by all!

We will be meeting in September/October, with the date and venue to be confirmed. Please contact me for further information about our group and we look forward to hearing from you, or better yet, come out for lunch and meet us. We are very flexible, we can meet at a restaurant or in my home in Kitchener. Until we meet, keep your smile brimming! :o) Betty

For more information, please contact Betty Van Luven: [bvanluven@rogers.com](mailto:bvanluven@rogers.com) or 519-954-3765

## WORLD WIDE WEB SITE VISITS

During the three months of April-June 2012 our web site logged a total of 6,584 visits, an average of 506 a week. The number of visitors may have been somewhat larger because of the way the information is compiled. There were no surprises in the way visitors found us (mainly through Google using predictable search terms) and the most frequently accessed pages (archived questions, Ask the Doc, the Society, research, and the Newsletter). More interestingly, visits originated from all over the world. Almost two thirds were from North America, as might be expected, with visits from the U.S. outnumbering those from Canada by more than 3:1. Visits from China accounted for one tenth of the total, although visits from Asia as a whole were half those from Europe. The main European countries of origin were the U.K., Germany and Ukraine. We even had a handful of visits from South America and Africa.



## CHEQUE PRESENTED TO LIVER CLINIC



On July 24, the Society's President, Barbara Badstober, presented a cheque for \$10,000 to Dr. Jenny Heathcote, leading researcher at the Francis Family Liver Clinic at Toronto Western Hospital, for use in research into PBC. Dr. Heathcote expressed her gratitude for the contribution and Josh Lai, of the Toronto General & Western Hospital Foundation explained that the hospital would be marking their appreciation by hanging a plaque recognizing the Society's cumulative contributions to the program.

## CANADA HELPS

**What We Do** CanadaHelps is a registered charity with a goal to make giving simple. Through CanadaHelps.org, anyone can donate online to any registered Canadian charity. We have proudly facilitated over \$100 million in charitable donations through our website since it was launched in 2000.

**For Donors** CanadaHelps is a one-stop-shop for giving. We made donating online easy and secure.



## CONTACT US

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[www.pbc-society.ca](http://www.pbc-society.ca)

## ONLINE VIA FACEBOOK

### Alberta North

[www.facebook.com/pages/Canadian-PBC-Society-Alberta-North-Region](http://www.facebook.com/pages/Canadian-PBC-Society-Alberta-North-Region)

### Ontario

[www.facebook.com/pages/Canadian-PBC-Society-Ontario-Region](http://www.facebook.com/pages/Canadian-PBC-Society-Ontario-Region)

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