



PBC connections

A Canadian PBC Society Newsletter

2015 • VOLUME ONE

APRIL 2015 ANNUAL GENERAL MEETING

On April 15, 2015 The Canadian PBC Society held its Annual General meeting in Toronto. There were eight people present. President, Barbara Badstober had received an additional 29 proxy votes, thus exceeding the required quorum of 15 outlined in the Society's constitution. At the meeting the 2014 minutes and financial statements were read and approved. Following Barbara's president's report, there was a lengthy discussion about the applicants for the 2015 Executive, which resulted in the following new appointments Gail Wright – President, Betty Van Luven-Vice President and Treasurer, Cheryl Watts – Director.

Barbara Badstober and Lenore Bennett agreed to continue as Directors providing continuity for the organization.

Introducing our newest members of the Canadian PBC Society Executive:



Gail Wright, diagnosed in 2012 with Stage 1 PBC, is excited to take on the role of President for the Canadian PBC Society. Organized, energetic and capable, she is committed to maintaining a strong organization to Reach for the Cure! Gail is married with three sons. She lives in Toronto.



Betty Van Luven hails from South western Ontario. Betty is a gifted speaker and dynamic leader. She has been active in the Canadian PBC Society since 2012 and has proven an invaluable support to all her PBC friends. Betty brings enthusiasm to her role of Vice President.



Cheryl Watts lives in Edmonton. She is an accomplished author and leadership consultant. She has written and published leadership books and has a Masters of Leadership from Royal Roads University. Cheryl has Stage 3 PBC and she recently completed a two year PBC study. Cheryl brings her communication and design skills to the Executive.

president's message



The rich colours of our Canadian fall make me grateful to live in such a wonderful country. How lucky we are. While taking comfort in autumn's warm reds, yellows and oranges, it occurred to me that the support and love we get are perhaps the key to our well being.

Support from health care professionals - we are especially thankful to those who have taken the time to see us as individuals, who speak to us directly and openly - sometimes there are not as many options as we would like but our health care professionals provide us with information and plans to go forward, and understand our frustrations. **RESPECT**

Support and understanding from our families and friends - whose love, smiles and kind gestures are so important. They are there with us and for us at our best and our worst. They understand when we've had a tough day. **COMFORT**

Support from our peers - to have a community of others who are going through the same things. We relax in the safety of knowing we are there for each other - chatting on Facebook, having lunches, planning fundraisers. **ACCEPTANCE**

Support from The Canadian PBC Society - that provides up to date and accurate information to PBCers and our health care providers and encourages personal support between our members. Equally important, we continue to reach for a cure by supporting research. **HOPE**

I ask you to consider each type of support you have received along the way and how that support has helped you on your journey. We all want and need: Respect, Comfort, Acceptance and Hope!

We count on your support, please renew your membership in The Canadian PBC Society today by completing and returning for form in this newsletter. If you are not already a member please join us now.

Warmest Regards,

Gail Wright

President, Canadian PBC Society

CANADIAN PBC SOCIETY NEWS

The 2015-2016 Executive is set to take on many new challenges, leading The Canadian PBC Society into the future. Along with the transition of the day to day operation, from the old executive to the new, they are very busy working on updating the PBC brochure in both French and English as well as looking at how to improve the PBC website: www.pbc-society.ca

Fundraising events in the Toronto area are being planned including: A Christmas Fundraiser in November, 2015 and A Day at the Races in May 2016 (back by popular demand).

It is wonderful to know that many regional groups are still active, supporting each other and having fun! Please keep sending updates and photos!

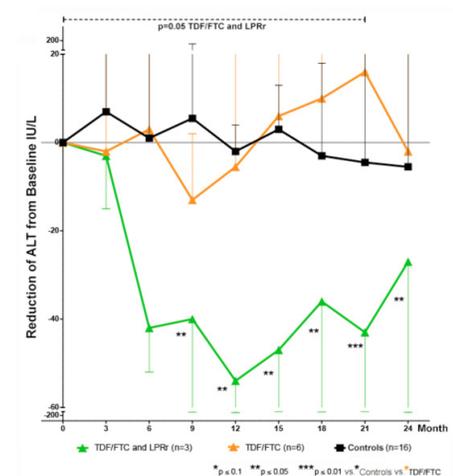
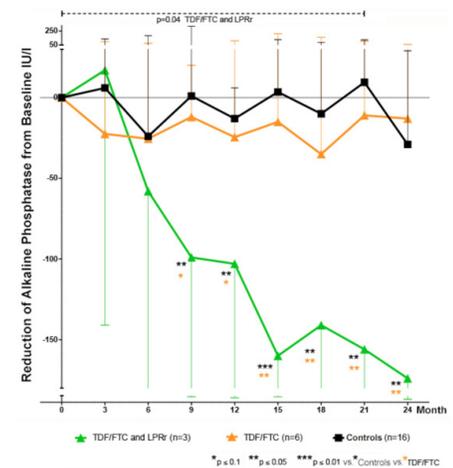
RESEARCH UPDATE FROM DR. ANDREW MASON

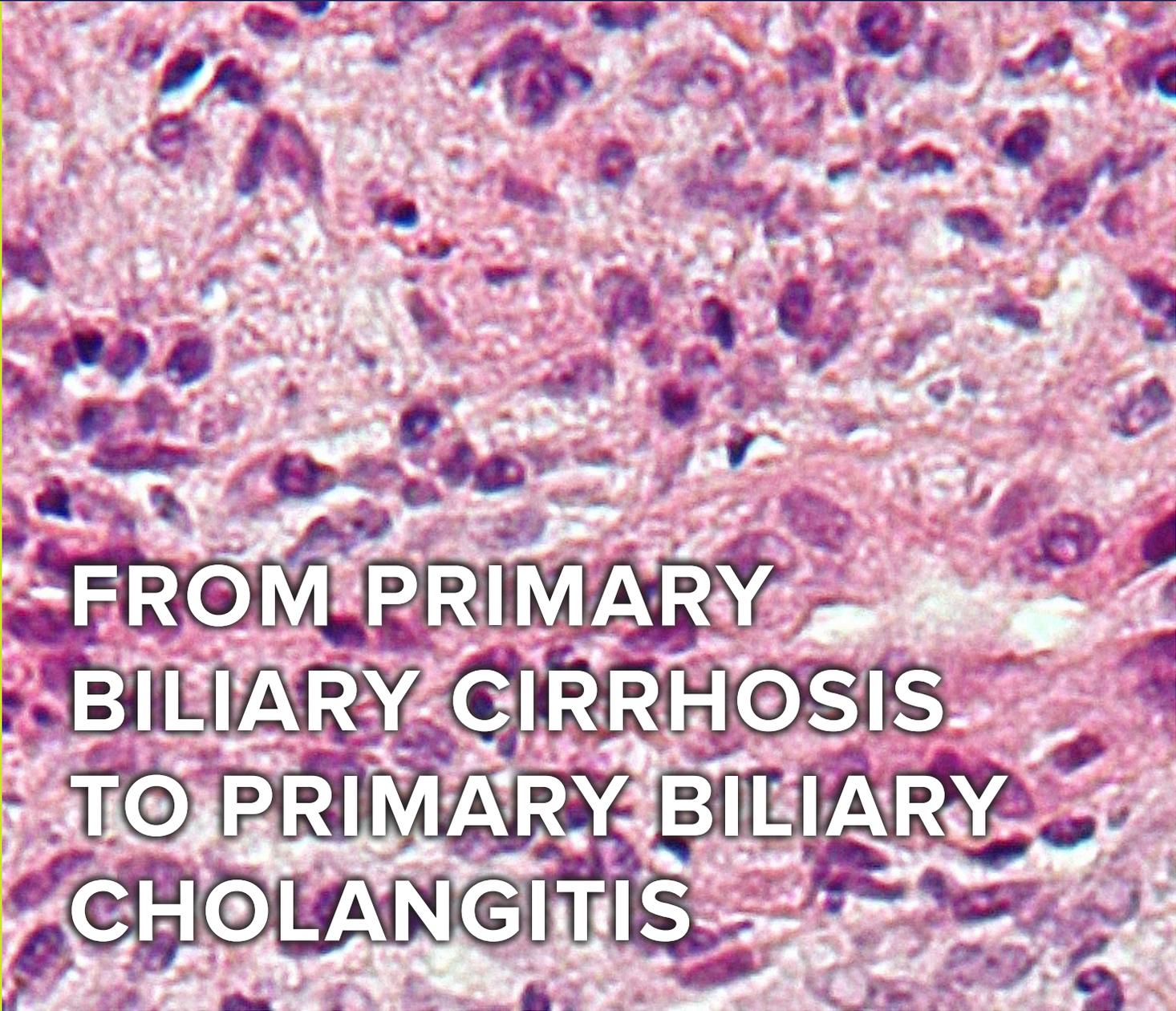
My laboratory has been studying a human betaretrovirus that we characterized in patients with primary biliary cholangitis (PBC) and used antiviral therapy to treat disease in clinical trials. Other researchers have published an article showing that combination protease inhibitors (lopinavir/ritonavir) and reverse transcriptase inhibitors (tenofovir/emtricitabine) completely normalized liver tests within 12 months in a patient with HIV, human betaretrovirus infection and PBC (Lancet 2011). We have used animal models infected with the same virus and found that these treatments can eliminate cholangitis.

Recently, we conducted a randomized controlled trial using the same anti-retroviral treatment in PBC patients who were unresponsive to ursodeoxy cholic acid and offered patients the treatment for 24 months. The enrollment was limited, as the majority of patients could not tolerate lopinavir/ritonavir, protease inhibitors. Ultimately only 3 patients remained on full treatment (tenofovir/emtricitabine and lopinavir/ritonavir) for 24 months, whereas 6 remained on tenofovir/emtricitabine alone. We used 19 clinic patients as a comparison group.

Patients on tenofovir/emtricitabine alone demonstrated comparable changes in their liver tests as the historical control group of clinic patients. Whereas patients on full treatment with tenofovir/emtricitabine and lopinavir/ritonavir experienced a significant reduction in the biliary cell marker, alkaline phosphatase, and the liver cell marker, alanine aminotransferase (Figure). All patients on antiviral therapy experienced improvement in symptom scores.

These studies are the first to show a clinically meaningful improvement in liver tests as well as improvement in symptoms using antiviral therapy. However, the frequency of side effects from lopinavir/ritonavir is more than double reported for HIV and better tolerated combinations of antiretroviral therapy will be required for future studies.





FROM PRIMARY BILIARY CIRRHOSIS TO PRIMARY BILIARY CHOLANGITIS

As some of you may have heard, PBC has had a name change, from Primary Biliary Cirrhosis to Primary Biliary Cholangitis. In June 2015 the following announcement was made by Linie Moore of the PBCers Organization:

“After a year of discussions, debates, meetings and numerous e-mails, the Primary Biliary Cirrhosis name change was approved last month by the AASLD (American Association for the Study of Liver Diseases) and EASL (European Association for the Study of the Liver). It is a privilege to announce effective June 2015, Primary Biliary Cirrhosis will be known as Primary Biliary Cholangitis. This is a worldwide change, not just US.”

Overall reactions to the name change in the PBC community have been positive.

Time to make the change from 'primary biliary cirrhosis' to 'primary biliary cholangitis'

Angela C Cheung MD FRCPC¹, Aldo Montano-Loza MD MSc PhD², Mark Swain MD MSc FRCPC FAASLD³, Catherine Vincent MD FRCPC⁴, Eberhard Renner MD FRCPC⁵, Morris Sherman PhD MB FRCPC⁶, Harry LA Janssen PhD MD^{1,7}, Andrew L Mason MBBS FRCPI²

The prognosis of patients with primary biliary cirrhosis (PBC) has improved substantially since the disease was first described >150 years ago by Addison and Gull (1). Over the past three decades, the improvement in transplant-free survival has been mainly driven by the widespread use of ursodeoxycholic acid (UDCA) and the timely detection of disease (2). The moniker 'primary biliary cirrhosis' was adopted in 1950 (3). However, it has been subsequently acknowledged that the use of the term 'cirrhosis' is a misnomer in patients presenting with early stage disease and histological evidence of chronic nonsuppurative destructive cholangitis (4). In early reports, a significant proportion of PBC patients presented with jaundice and decompensated cirrhosis, and virtually all of these patients died from cirrhosis and liver failure within 10 years of their diagnosis (3,5). Now, <50% of patients are documented to develop cirrhosis and the median transplant-free survival of UDCA treated patients is >20 years (2).

Because many patients with PBC do not have and will never develop cirrhosis, this label has understandably upset many patients with PBC, who have now pushed for a change. During the European Association for the Study of Liver Monothematic Conference on Primary Biliary Cirrhosis in May 2014, representatives of patient advocacy groups initiated discussions with hepatologists to change the eponym 'primary biliary cirrhosis' to one that would more accurately reflect the features of the disease. From the patient perspective, the term 'cirrhosis' is misleading on several fronts, and may lead to stigmatization and confusion with alcohol-induced cirrhosis, as well as a lack of clarity concerning the stage of disease and the prognosis. This initiative for a name change was supported by patients around the globe, including members of the PBC Foundation (United Kingdom), the PBCers (United States) and the PBC Society (Canada).

Over the past year, patients and physicians have worked together to achieve a consensus regarding the name change. In a poll involving more than 50 hepatologists from around the world, 95% agreed that the name should be updated, and >80% believed that it would be best to maintain the acronym 'PBC'. Similarly, the vast majority of >1000 patients surveyed supported the name change. Several terms were considered to replace 'cirrhosis', including 'cholestasis' and 'cholangiopathy'. The final consensus was that 'primary biliary cholangitis' provided the most accurate description of the disease, echoing the histological description first coined by Rubin et al (4) in 1965.

Understandably, there is concern that the terms 'primary biliary cholangitis' and 'primary sclerosing cholangitis' (PSC) are now fairly similar, differing in acronym by only a single letter. This may lead to confusion and misinformation, given that patients with either disease may develop biochemical evidence of cholestasis and symptoms of fatigue, pruritus and jaundice. Despite some similarities in presentation, however, there are currently substantial differences in available treatments and prognosis. It is, therefore, imperative that the medical

community strives to minimize inaccuracies and misconceptions that may develop from this initiative.

While this change in terminology more accurately reflects the natural history of PBC in the current era, it also highlights the knowledge gaps in our understanding of PBC. In fact, some hepatologists have raised the concern that 'primary biliary cholangitis' is a non-specific moniker that underscores our lack of understanding of the etiology and pathogenesis of PBC. The risk factors that trigger different phenotypic manifestations of PBC and the response to UDCA therapy are largely unknown. Genome-wide association studies have demonstrated a strong association with human leukocyte antigen alleles and multiple single nucleotide polymorphisms associated with genes along the interleukin-12 axis (6,7). However, the role that specific risk alleles play in PBC has yet to be defined and no risk loci specific to PBC have been identified (7). Whereas several environmental factors have been linked with PBC, including xenobiotics and infectious agents such as proteobacteria and retroviruses, none have categorically been associated with the development of disease (2,7). Clearly, a better grasp of the disease process would lead to a more appropriate name for PBC.

Despite these concerns, it is timely that the Canadian Association for the Study of the Liver and the Canadian Liver Foundation formally recognize the change from 'primary biliary cirrhosis' to 'primary biliary cholangitis' in conjunction with the American Association for the Study of Liver Diseases and the European Association for the Study of the Liver. While this revised terminology is, in part, a reflection of our patients' need for more clarity in understanding PBC, we hope that it will also usher in a revolution in the research and management of primary biliary cholangitis.

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regional connections

ALBERTA NORTH

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ATLANTIC

We had a great turnout for our spring get-together in June. There were 14 of us – 10 who belong to the PBC Group, two husbands and a sister. It was so nice to see some of the ladies that we haven't seen in a long time.

Five of us also had a lovely get-together on October 28. We enjoyed a delicious lunch and lively conversation. Judi, our president, gave each of us a beautiful book mark that she had made. We are sorry that more of you were not available, but hope to see you in May 2016.

Stay well and have a wonderful holiday season.

or more information, please contact **Judi Pemberton:**
atlantic@pbc-society.ca or 902-798-5554

BRITISH COLUMBIA

For information, please contact **Kathryn Swift:**
bc@pbc-society.ca

GOLDEN HORSESHOE REGION

For information, please contact **Karen Isbister,**
kisbister@cogeco.ca or 905-336-3502, or **Jackie Gay,**
gingerjack@cogeco.ca or 905-937-1081.



MANITOBA

The Manitoba Chapter of the PBC Support Group met for dinner on June 25 at Tony Roma's Restaurant. There were 13 in attendance, almost a record for us. As well as members there were 3 spouses and one person brought her daughter. We had a great meal and welcomed some new friends for the first time. Conversations were animated and stories shared.

One of our members, Karen L. has set up a private local Facebook site for us called PBC Winnipeg and Area Support Group. At least 2 of our members are awaiting transplants in the near future. The meeting planned for late September was cancelled. I hope to arrange something in the next few weeks.

For information, please contact **Carol Seburn:** Manitoba@pbc-society.ca or 204-254-5226

TORONTO

For information, please contact: info@pbc-society.ca

SOUTHWESTERN ONTARIO

We held a get-together in June at Moxie's Restaurant in Yorkdale Plaza. A great time was had by all. Gail Wright, our new President, was able to join us.

Although I am the coordinator for Southwestern Ontario, I also arrange and participate in get-togethers in the GTA, Hamilton and Burlington areas. I encourage members to contact me if they would like to participate in an informal get-together. Did I mention that food is involved?

I am looking to set up a get-together, before the snow flies, in the Kitchener/Stratford area, which is pretty close for the London members. Please contact me at the address below.

For more information, please contact **Betty Van Luven:** ivanluven@rogers.com or 226-339-3173



OTTAWA

For information, please contact **France Foucault**:
ffoucault@hotmail.com

QUÉBEC

La fin de semaine du 18 et 19 septembre 2015, le Groupe de Québec/Ontario s'est réuni à l'Auberge des Quatre matins à Saint-Côme située en plein coeur de Lanaudière. Cette année le caractère de la rencontre se voulait décontracté, amical et relaxant.



Les membres ont donc bénéficié de ces deux journées pour échanger librement sur la CBP, leur

expérience et les bons trucs découverts à travers le temps. Nous avons eu le privilège de nous connaître et de nous apprécier davantage. Outre les enrichissantes conversations, les bons fous rires, de savoureux repas ont été servis. Les bains nordiques et massages faisaient partie de nos divertissements.

Le groupe de personnes présentes s'entendent à dire qu'une rencontre similaire agrémentée d'une conférence serait fort appréciée l'année prochaine. La région de La Malbaie a été ciblée comme lieu de rencontre, nous y travaillons déjà. J'invite les personnes intéressées à se joindre au groupe, à me contacter, vous en sortirez grandies. Merci aux membres de faire partie du Groupe, de participer aux activités et d'aider d'autres personnes à mieux vivre avec la CBP.

For more information, please contact **Francine Lamontagne**, Représentante Québec, Société canadienne de la PBC:
quebecrep@pbc-society.ca

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.

ONLINE VIA FACEBOOK

Alberta North

www.facebook.com/pages/Canadian-PBC-Society-Alberta-North-Region

Ontario

www.facebook.com/pages/Canadian-PBC-Society-Ontario-Region

Quebec

www.facebook.com/pages/Société-canadienne-de-la-CBP

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