



A Canadian PBC Society Newsletter

2010 - VOLUME 2

## **THE ANNUAL GENERAL MEETING**

The Annual General Meeting of the Canadian PBC Society was held on Friday, May 7, 2010, in Toronto. Eight members and Randall Starr of Starr Celeste-Falconeri, Chartered Accountants LLP were in attendance and the President held 31 proxies.

Members passed a motion to amend the Society's by-laws to require the AGM to be held within 120 days after the end of the fiscal year (extended from 90 days). The amendment was required because of difficulty in obtaining audited financial statements within the 90 days.

The financial statements were presented by Randall Starr and accepted by the meeting. Randall Starr is stepping down from being the Society's auditor and the President thanked him for his work on our behalf.

The President reported on a successful year with increased membership, an updated web site with more visitors, and fundraising efforts that matched the previous year's despite a difficult financial climate.

The members approved by a unanimous show of hands the following slate of officers for 2010.

### **The 2009 Executive**

President .....Barbara Badstober  
Treasurer.....Open  
Recording Secretary .....Mickie Nikoras  
Membership Secretary .....Lenore Bennett  
Communications .....Alwyne Graham (Editor, PBC Newsletter)  
Corresponding Secretary .....Carol Pye  
Archivist: .....Esther Schwarz  
Fundraising.....Open

### **Regional Coordinators**

British Columbia .....Kathryn Swift  
Alberta .....Shauna Vander Well  
Manitoba.....Carol Seburn  
Greater Toronto .....Open  
Quebec .....Francine Lamontagne  
Atlantic .....Judy Pemberton

The President conveyed thanks to John Zeller, the Society's lawyer, for his ongoing contribution and to Katherine Fuerst for her help in arranging the meeting.



## FUNDRAISING

This year's **Day at the Races** was an unparalleled success, with perfect weather, a record number of people attending and a variety of donated items for the silent auction. Our young friend, J, once again sold her handcrafted jewelry pieces.

Everyone enjoyed a delicious buffet lunch and enthusiastically cheered on their chosen horses, or simply enjoyed watching and mixing. Children were formally invited for the first time, and the presence of a number of youngsters added to the general happy atmosphere. Dr. Gideon Hirschfield, accompanied by his little daughter, gave a brief speech pointing to the progress being made in research into PBC and thanking the Society for its help in recruiting members and providing funds for the data base.

Special thanks to everyone who donated an item for the silent auction, to Schonberg Farm for their continued support and sponsorship of a PBC Society race, to the Woodbine staff for their friendly and efficient service, and to Barbara Badstober for all her hard work.

***Don't miss next year's event!***  
***Mark your calendar for Sunday, July 12, 2011.***



## RESEARCH

### MUSCLE FUNCTION IN PBC: CLUES TO THE CAUSES AND TREATMENT OF FATIGUE?

**Prof. David E Jones & Prof. Julia L Newton,  
Newcastle University, UK**

Several studies from North America and Europe (including from Canada) have shown that PBC patients can be affected quite significantly by fatigue. It is very striking and perhaps surprising that the severity of this fatigue is not associated with how bad the underlying liver disease is, which means that treating the liver will not necessarily improve how people with PBC feel. As in any group of people (and it is important to remember that fatigue can affect lots of different groups of people) fatigue in PBC can be caused by many different things (and frequently lots of different little things combining). This is very important when thinking about how best to treat fatigue because one of the first things we do when seeing a fatigued PBC patient in the clinic and trying to improve the fatigue that they experience is to remember that PBC patients can suffer from other diseases (e.g. thyroid disease) or the affects of medication, which are potentially easily treated causes for fatigue.

Even when, however, such other fatigue associated disease factors are not present, PBC patients experience a particularly characteristic type of fatigue which is frequently likened to "batteries running down". We have shown that this type of fatigue, with its characteristic feelings and descriptions, is associated with reduced amounts of physical activity and with difficulties that people describe in repeating physical activities multiple times. This led us to becoming interested in whether or not there really is a problem with muscle function in PBC. For many years it was very difficult to study this until technology led to advances in imaging techniques that allowed us to use magnetic resonance imaging (or MRI; a technique familiar to many people as a form of body scanning). MRI has allowed us to study what is happening within



muscle to see whether or not they are functioning normally in people who perceive themselves to be tired. This work has now been published in three different scientific papers and provides really important insights in to what might be happening to PBC patients with fatigue and, importantly, what we could do about it.

Our initial finding was that PBC patients as a group have a tendency to have too much acid in their muscle at the end of a fixed period of exercise. In this sense PBC patients are very similar to people who have done an extreme amount of exercise. The amount of exercise that PBC patients did in the study to get this effect was, however, minimal. A second effect was also noted that this time was associated with fatigue. This was that PBC patients had a variable capacity to recover from excess acid in the muscle. People who were able to recover very quickly did not have fatigue, regardless of how much acid there was in the muscle to start with. People who had elevated levels of acid, and who couldn't recover quickly, were typically very fatigued. A second study showed that similar changes in how muscles power themselves were also present in heart muscle suggesting that this is an effect present throughout the body in PBC not just restricted to one particular set of muscles. Our most recent study went on to look at what happened when people underwent repeated exercise with a recovery period in between. What this study showed is that, perhaps surprisingly, PBC patients were quite normal in their ability to reduce acid production in their muscles with repeat exercise (when normal people exercise followed by a period of rest and repeat exercise they produced less acid the second time

they exercised because their body was prepared). PBC patients had, in fact, a striking ability to increase their capacity to handle acid. This is an important finding because it suggests that whatever is happening in the muscles of PBC patients it is potentially reversible. What was very striking was that when people went on to exercise for a third time improvement following exercise was lost and, in fact, patients typically started to take longer to recover. There was a striking difference between the fatigued and non-fatigued PBC patients, with fatigued PBC patients in particular showing marked worsening of their capacity to recover.

What does this all therefore mean to a patient with PBC who feels their muscles are tired? What it suggests is that there is probably a PBC-specific abnormality in how muscles produce acid and there is a secondary process which represents how the body's normal processes of recovery can handle this. Some people are good at this and some people are less good and the less good people are more fatigued. Importantly, all the PBC patients we have studied to date improve with certain types of exercise in terms of how their muscles handle acid it's just that they don't improve enough. There is, therefore, a capacity issue which we need to improve on. The obvious question is how post-exercise acid recovery can be improved in PBC patients, and the simple answer could be exercise training as improvement in the handling of acid is what underpins athletic training regimes. The final part of our study was therefore a small study to look at how well patients could manage exercise training and whether it improved its symptoms. The answer is that it was well tolerated and the symptoms improved. It is important now that this work be built on and repeated by other labs and we need to work out what is the right exercise programme to give people the most benefit and no harm. So at the moment we would not recommend exercise to everybody and suggest people taking exercise needs to consult with their doctors as whether it is safe in their own personal situation. The important message is, however, that there may be a specific and treatable muscle component to fatigue in PBC and that exercise training is a very attractive direction for treatment where studies are badly needed.

## **Update from the Liver Clinic, Toronto Western Hospital**

### **Research**

The Summer of 2010 has been anything but quiet for the autoimmune liver disease programme at Toronto Western Hospital. Clinics are bustling as words spreads across Ontario of its particular interest in managing patients with primary biliary cirrhosis, primary sclerosing cholangitis, autoimmune hepatitis and autoimmune pancreatitis. With the ongoing support of its patients, and its local, national and international collaborators, important breakthroughs continue to be published. As reported online this month in Nature Genetics (<http://www.nature.com/ng/journal/vaop/ncurrent/abs/ng.631.html> and <http://www.nature.com/ng/journal/vaop/ncurrent/abs/ng.627.html>), the group has gone on to discover more genes that predispose patients to PBC. These genes are already known as risk factors for other autoimmune diseases (such as lupus, colitis, and asthma) and so continue to uncover the web of genetic risk that means a patient becomes ill with PBC.

Future plans include new drug trials for patients with autoimmune liver disease, PBC in particular. Dr. Hirschfield is also working closely with the Mayo Clinic to help patients with the related bile duct disease primary sclerosing cholangitis. Similar breakthroughs are hoped for this disease in the years to come.

For an update on Dr. Hirschfields' latest publications follow the link <http://www.ncbi.nlm.nih.gov/pubmed/?term=Hirschfield+GM> or for more information visit our website, <http://www.torontoliver.ca/>, or email us at [aildwh@gmail.com](mailto:aildwh@gmail.com)

### **Database**

The database to explore how PBC begins and develops has been expanded with the completion of collection of clinical data from Calgary and Edmonton.

Phase 3 Clinical Trial of the new Intercept drug will be coming in January 2011.

### **Communication**

New patient education material and an updated Liver Centre website will be coming soon.

## **REGIONAL NEWS**

### **ATLANTIC**

Seven of us enjoyed a social meeting on June 17. The Bingo Bowling was fun, and it was great to get together with some of the group. Margaret Hemeon joined us, the first time she was able to be with us in several years. Six of us bowled, and Mona had the difficult task of keeping track of all our gutter balls. We hope to see everybody in September.

**Contact Judi Pemberton at [atlantic@pbc-society.ca](mailto:atlantic@pbc-society.ca) or 902-798-5554**

### **QUEBEC**

Members of the Quebec group maintain communication via the Internet. Since members live in different regions of Quebec, meetings are difficult to hold. However, we are trying to organize an informal meeting at the end of the summer, date to be confirmed.

Les membres du Groupe du Québec maintiennent une communication via Internet. Étant donné que les membres habitent différentes régions du Québec, les rencontres sont difficiles à tenir. Toutefois, nous tentons d'organiser une rencontre informelle à la fin de l'été. La date reste à confirmer.

**Contact Francine Lamontagne at [QuebecRep@pbc-society.ca](mailto:QuebecRep@pbc-society.ca)**

### **TORONTO & GTA**

**Dr. Hirschfield will be addressing a meeting on October 2, 2010.**

Location: All Purpose Room. 2, Covington Road. Toronto.

Time: 10:30 am. Light refreshments available.

**Contact Barbara Badstober at [info@pbc-society.ca](mailto:info@pbc-society.ca) or 1-866-441-3643**

### **ALBERTA NORTH**

Eight of us had a coffee meeting in June, when we chatted about drugs and vitamins (some people were aware that ADEK vitamins are available while others thought they were permanently unavailable) and compared stories about itching and fatigue. There was unanimous support for a fourth annual barbecue to be held on September 18 at Gary and Soule Russel's house. There will be a small silent auction and we're hoping for a big turnout!

**Contact Shauna Vander Well at [AlbertaN@pbc-society.ca](mailto:AlbertaN@pbc-society.ca) or 780-962-6217**

### **ALBERTA SOUTH**

Unfortunately, it has not been possible to get a group off the ground, despite having 12 people interested and in email contact.

### **MANITOBA**

Small meetings of 3 or 4 people were held in April and on May 27. Others are planned.

**Contact Carol Seburn at [manitoba@pbc-society.ca](mailto:manitoba@pbc-society.ca) or 204-254-5226**