

Fall, 2005



Iron Filings

From Super Woman to a Premature Retirement

In the early 1990s, I was feeling the best I ever had in my whole life. Even though I had recurrences of sickness at different times in my life, I was feeling physically fit and proud to hang on to what had proved to be a difficult working position. I was in my early 40s and a mom of two teenagers who had started university. I was an office manager in a pretty important organization, and I was an athlete, running 10 km races in the summer and training at the gym while running the rest of the year. I was also a dedicated volunteer for the Children's Hospital.

In March 1995, at 43 years old, I was diagnosed with hypothyroidism. The normal treatment of thyrosine helped me with some of the symptoms, but I kept having constant

infections, swelling of the lower abdomen and legs, very dry skin, low energy and a very painful hip. Nothing was showing in the different tests my doctor requested other than a bit of low hemoglobin, which was treated with iron supplements. I also had occasional severe pain in my upper right abdomen during this period.

In February, 1998, a minor car accident in which I hurt my abdomen muscles caused more problems than anticipated at first. A few months later, I found I was always tired: I had a constant headache, I was nauseous most of the time and my muscles felt as if they were getting shorter every day. Even getting dressed in the morning became very difficult. I also had a lot of swelling of my lower body and legs. I was a mess.



Member in the Spotlight, Thérèse Dupuis

In June 1998, my doctor told me that she wouldn't put me on a leave of work for more than three days because it wouldn't do me any good. In September, still in pain and with no relief in sight, I handed in my resignation from work. Tests revealed nothing more than borderline anemia, PMS and depression. My

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The Newsletter of the
Canadian Hemochromatosis
Society

HEMOCHROMATOSIS

Very common • Virtually unknown • Potentially fatal • Easily treatable

What is it?

The excess storage of iron in the body.

What is the cause?

Primarily hereditary.

Most common symptoms

Chronic fatigue, joint pain, irregular heart beat, mood swings, confusion, bronzing of the skin, loss of libido and abdominal pain.

Most common complications

Liver and heart disease, diabetes, arthritis and hormonal irregularities.

Tests required for diagnosis

Serum ferritin, transferrin saturation percentage and genetic testing.

Treatment

Phlebotomy treatments (bloodletting) which are ongoing for life.

Reference reading

The Bronze Killer; Ironic Health; The Iron Elephant; Iron Disorders Institute Guide to Hemochromatosis.

Changes Ahead for CHS

A couple of days ago I received two of my favourite magazines in the mail and noticed that they have both been redesigned. There must be something in the air because, as you'll notice, *Iron Filings* has once again been given a facelift. It's part of a re-branding initiative that includes a major updating of our website – although the launch is still a few weeks away – and a new logo that reflects the genetic aspect of hemochromatosis.

We hope these changes will improve our service to our members, and help promote the Society and its goals to a wider audience.

This activity points out, again, how few of us there are to do the work of running the Society and how much money and time

it all takes. We are

coming up to the 25th anniversary of CHS in 2007 and we look with pride to the advances we've made in those years. At the same time we are frustrated by our lack of headway in achieving the finan-

cial stability that would let us continue to employ professionals to help us keep moving forward. If we have learned anything since our founder, Marie Warder, had to step down because the burden of running the Society was destroying her health, it's that we can't do it all ourselves. Small health charities like ours must employ professionals or be swept away by the competition.

Our fundraising results have been disastrously low this year, partly because world catastrophes have tugged on the purse strings of potential donors and partly because we have yet to launch a truly effective campaign. Without adequate fundraising, CHS faces a bleak future and our mission – to end needless suffering and early death from hemochromatosis in Canada – will not be fulfilled.

To begin addressing these realities, you will read about our new membership drive and a piece on planned giving from our fundrais-

ing committee.

Our member in the spotlight this issue once again exemplifies the kind of committed, passionate individuals we have within our membership and we hope this inspires you to think of new ways you can contribute. We also appeal to you to spread the word about CHS to your family, friends and neighbours and encourage them to become members.

Whenever I get discouraged and wonder if we are making a difference, I go to our



Without adequate fundraising, CHS faces a bleak future, and our mission – to end needless suffering and early death from hemochromatosis in Canada – will not be fulfilled.

newsboard and read the stories of people and their families affected by hemochromatosis, desperately looking for help.

I also use the newsboard as a barometer in choosing topics to cover in the newsletter. It's gratifying to hear that someone has been di-

agnosed because of information they received from us, but it's easy to forget that diagnosis is just the beginning of a difficult journey. The many newsboard questions about treatment told me it was time for some basic information again. The protocol for treatment has evolved and a piece on phlebotomy therapy is included here from our very own Dr. Sigfried Erb, Internal Medicine specialist at VGH, based at UBC.

We also have a piece from Dr. Sam Krikler, a hematologist and former board member. He has filed a report from his trip to the BioIron Conference in Prague this past May.

Finally, this issue includes our thanks to our donors, without whom none of our vital work would be possible.

We hope you enjoy your read. When you're finished, pass your copy along to a friend.

Every little bit helps in spreading the word.

Elizabeth Minish, President

IRON FILINGS

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The Disorder

Hemochromatosis is the most common genetic disorder afflicting Canadians. It is a crippling, potentially fatal condition caused by iron overload in organs, joints and tissues. The complications caused by the disorder are preventable.

Our Purpose

The society is dedicated to preventing the unnecessary suffering and death caused by hemochromatosis by promoting awareness and early diagnosis while supporting those affected by the disorder.

Thérèse Dupuis
Member in the Spotlight
continued from page 1

doctor prescribed anti-depressants, which I never took. Of course, surrendering to the pain wasn't the right thing to do financially, and since I didn't have a diagnosis of anything serious, I lost any chance on my disability insurance benefits.

After a few weeks of rest from work, some symptoms began to disappear. I kept on jogging and stretching as much as I could, and I found that if I stopped completely for more than a couple of weeks, I developed high blood sugar. So I kept on working out. The stretching proved to be essential to help with the stiffness, and the jogging, even very limited, kept me feeling good mentally.

I tried to go back to a part time job, but after about a month I started to be very tired and the symptoms came back. Since then, I have to calculate very meticulously the amount of energy I spend. I started my own office services business in spring 2001, so when I have work to do, I organize my energy limit around it.

In spring 2001, I was diagnosed with chronic fatigue with fibromyalgia symptoms. An infectious disease consultant prescribed a muscle relaxant. He said that it would help the swelling. It was working but I still had to be careful since fatigue was the culprit.

Shortly after that, one of my sisters was diagnosed with a very elevated iron level. It was done in a routine test by the only doctor around here that I think checks for iron-overload. At the news, my sister was the joke of the family since all the other girls were or had been treated for iron deficiency anemia. But we were informed that we should all be checked for HH.

After brief research, I knew I had it. I had so many of the symptoms. At first, I was told that my iron level was not high and that I would be checked again in three or four years. I requested a genetic test, but was told everything was good. After a few months and more research, I asked for a copy of the genetic test. The report stated very clearly that I was homozygous for the mutation C282Y and that I should see a specialist a.s.a.p.

Later on, a liver biopsy showed that my

liver had a dangerous iron concentration. My specialist seemed to be in disbelief that there was no tissue damage. The phlebotomies were started right away. From October 2002 to January 2003 I had 15 phlebotomies. Since then, I have been taking care of the phlebotomies myself. With a note from the doctor, I go to the Red Cross every three months.

My mother died at age 74 of diabetes and had many other symptoms of HH. My family is now convinced that she had HH. My father is almost 97 year old and is suffering only mild dementia. Only three girls of a family of 12 are homozygous for the mutation C282Y. Others have only one mutation even though quite a few nieces and nephews are affected.

I still exercise, dance and work a bit with my company, so I consider myself lucky. My energy level has been good since the phlebotomies, but I have to be very careful. Overdoing it can mean a week or two of a recurrence of extreme fatigue, swelling and stiffness.

I'm not dreaming anymore of doing 10 km races. My last one was in 1997. But at least I know where I stand. I don't have a real job and I can't be sure that I'll be able

to dance or jog tomorrow, but I know I have hemochromatosis. My last tests results were good and I know I was saved from almost certain liver disease thanks to the vigilant doctor who diagnosed my sister. Thanks to him, this May, my husband and I are going to visit our soon-to-be-doctor son in Scotland. I can also look forward to meeting our first grandchild who our daughter is expecting in June.

I'm a determined person when something has to be done. However, a lot of people are not able to act in their own interest. Even the ones who are informed about the existence and the symptoms of HH have to get information through their family physician and know how to request and read their own test reports. It is crucial that the facts about HH

get through to the public and to doctors to change this situation.

Life is good with or without limitations when you know and you accept what you're dealing with.

Therese was the first person to respond to our membership committee's new initiative when they put out the call for Regional Co-ordinators. She is also involved in the French version of our website.

My last tests results were good and I know I was saved from almost certain liver disease thanks to the vigilant doctor who diagnosed my sister.

Regional Coordinators – A New Initiative

Dr. Krikler's report from the International Biolron Society Conference (see page 7), tells us about the potential for international cooperation with our counterparts in the European Union. It's wonderful to know that awareness of hemochromatosis is growing around the world. Using "think globally, act locally" as a guide, our challenge is to be more effective at the community level, especially since we have only one office to serve the entire country.

To this end, the membership committee came up with the idea of creating a Regional Coordinator level to CHS. We hope to tap the energy, creativity and passion that exists in our membership to accomplish this goal. Regional Coordinators will create more support for our current contacts, and spawn new ideas and activity in the areas of awareness and fundraising that are more locally based.

Already, a dozen people from across the country have stepped forward, but we need to hear from more of you to make the program really effective. If you would like more information on becoming a Regional Coordinator, please contact the office at office@cdnhemochromatosis.ca.

Treatment of Iron Overload

A Hemochromatosis Primer

For newly diagnosed hemochromatosis patients, the process that leads to successful management can be bewildering. They often do not receive much information about what happens next and why it is vitally important to stay on top of their treatment. To address this, we have asked Dr. Siegfried Erb, a Vancouver-based liver specialist who treats many hemochromatosis patients, to answer some of the key questions relating to the treatment of iron overload.

Iron Filings: *What is the most important aspect of treatment that a person, newly diagnosed with hemochromatosis, should know?*

Dr. Erb: Many aspects of treatment are important, including management of complications, screening for liver cancer, avoidance of supplemental iron and appropriate vaccinations for hepatitis A and B, but, an aggressive de-ironing protocol is foremost.

IF: *How is this accomplished?*

Dr. Erb: We get rid of the excess iron by phlebotomy, which is the drawing off of a unit of blood, using the same procedure as a blood donation, but with much higher frequency.

IF: *Why does phlebotomy therapy work?*

Dr. Erb: Under normal circumstances, the body only absorbs about 10 per cent of the iron it takes in on a daily basis. The remaining 90 per cent is excreted. With Type I hemochromatosis, the most common form (See Spring 2005 for an update on types I to IV hemochromatosis.), which results from a mutation of the HFE gene, iron absorption is three to four times greater than normal. The majority of iron in the body is contained in hemoglobin, the oxygen-carrying protein in red blood cells. The rest is distributed in various iron transporting and storage proteins, and in bone marrow where red blood cells are produced. With iron overload, the excess iron, builds up over a few decades in inappropriate places, like the liver, heart and endocrine glands, where it causes problems like cirrhosis of the liver, cardiomyopathy, diabetes, arthritis, etc. Phlebotomies pull the iron out of these places and back into the red blood cells, which can be removed over and over again, keeping the hemoglobin stable as the patient gets de-ironed.

IF: *What is the benefit to being aggressively de-ironed?*

Dr. Erb: Studies clearly show that both survival rates and quality of life are significantly improved, even for patients who have already sustained organ damage. For those diagnosed early, before clinical symptoms are manifest, or there is evidence of liver dysfunction or cirrhosis, longevity is statistically the same as for the average person without hemochromatosis, in the absence of other risk factors for liver disease, such as alcoholism or hepatitis C.

IF: *If damage has already occurred, is it reversible?*

Dr. Erb: Some liver function can be improved, but not cirrhosis. There is a mild improvement in diabetes and heart function. Improvement in the joint symptoms is variable and there seems to be no improvement in some of the

hormonal complications, such as impotence. Even if there is no improvement, preventing progression of the disease process in damaged organs makes treatment worthwhile.

IF: *If it takes a few decades for the excess iron to build up and cause symptoms, why does it matter how quickly one gets rid of the iron?*

Dr. Erb: Firstly, low levels of iron overload cause fewer problems than higher levels. By the time a patient is clinically unwell, he or she is already at that critical point and it becomes imperative to get rid of the iron as quickly as possible. Secondly, while it is easy to mobilize iron from the liver, and liver cells turn over relatively frequently allowing the liver a great propensity to regenerate, the same is not true for the heart or other organs. It is important to maintain a very aggressive approach, at least for the first two years, to avoid further possible damage to the heart.

IF: *What do you mean when you talk about aggressive de-ironing?*

Dr. Erb: Iron can be mobilized out of the liver at 130 mg/day, or 900 mg/week. Since one unit of blood, 450 cc, contains approximately 225 mg of iron, one could theoretically remove four units of blood a week without

Glossary

Anemia – a blood condition involving too few red blood cells, insufficient hemoglobin or low volume. Iron deficiency is only one of many causes.

Ferritin – a protein that stores iron. It is produced by most cells in the body to bind iron until it is needed. As iron increases, more ferritin is produced to try to contain it. The body also uses ferritin to withhold iron from invading infections or cancer which need iron to thrive. Because elevated ferritin levels result from many causes, it is unreliable as the sole indicator of iron overload.

Hemoglobin – the oxygen carrying red pigment that gives red blood cells their colour. Contains most of the body's iron in normal circumstances.

Hematocrit – the percentage by volume that the red blood cells make up in whole blood. This measure is affected both by size and number of red blood cells. The other components of whole blood are white blood cells

and plasma. This measurement is used more in the United States than in Canada.

Macrophage – a category of cell widely distributed in the body, part of the immune response. They mount a defense by engulfing invading foreign cells.

MCV – Mean Corpuscular Volume. This measures the size of the red blood cells. Too small may indicate iron deficiency.

Serum iron – the amount of iron bound to transferrin in the blood.

TIBC – Total Iron Binding Capacity or total amount of iron that would be required to have every molecule of transferrin bound.

Transferrin – a protein that transports iron in the bloodstream. Normally about 1/3 of transferrin molecules have iron bound to them.

Transferrin saturation percentage – calculated by dividing the serum iron level by the TIBC. Eg., If serum iron is 150 and the TIBC is 175, then the transferrin saturation percentage is 86%.

exceeding the maximum rate of mobilizing iron out of the liver. As I said before, the liver has a rapid turnover of cells, and moving iron in and out is a normal function. Incorporating that iron into red blood cells is slightly slower. During the de-ironing phase, the goal therefore is to maximize the output of red blood cells by the bone marrow. A mild to moderate anemia will increase the bone marrow's output by 500 to 600 per cent. A normal red blood cell lives for 100 to 120 days. The average blood volume is 4.5 litres, or 10 units of blood. One unit therefore turns over every 11 days, 110 days survival/10 units of blood. Since the bone marrow can produce five to six times that rate with an anemia, it can therefore turn out a unit of blood every two days, 11days/5-6x-rate. Therefore, one could actually do a phlebotomy three to four times a week. Some patients who are extremely iron overloaded will require the placement of a venous access device to achieve this goal.

IF: That does sound extreme. What tests do you use to establish the optimum frequency for your patients during this phase?

Dr. Erb: During the de-ironing phase, we simply do a hemoglobin test on the drawn blood. By taking it from the bag, it avoids an extra poke for the patient. We start with weekly phlebotomies. Each week, the hemoglobin from the previous week is available. In this de-ironing phase, we want a mild to moderate anemia, 100 to 120 gm/L for men, and 90 to 110 for women. At this hemoglobin level, the bone marrow output will be at the maximum. After six weeks, if the hemoglobin is still greater than 120 (110 for women), we increase the phlebotomies to twice a week. After a further six weeks, if the hemoglobin is still too high, we could increase to three phlebotomies a week, if the patient will tolerate it, and doesn't have a problem with his veins. The average man with hemochromatosis requires 68 phlebotomies, the average woman requires 25. The average man with hemochromatosis could be completely de-ironed in as little as 30 weeks.

IF: How do you know when to stop the de-ironing phase?

Dr. Erb: Since we can't do phlebotomies faster than we can mobilize iron from the liver,

as we are limited by the output of the bone marrow which has very limited stores of iron, when all of the iron has been removed from the liver, the patient will develop a progressive iron deficiency anemia. We stop when the hemoglobin is less than 100 (90 in a woman). Alternatively, the patient will become symptomatic with fatigue and malaise. These symptoms are not related to the anemia, as one could easily withstand a hemoglobin much lower. Rather, they are related to the iron deficiency, as iron is required as a cofactor in many of the body's biochemical reactions. Once we think someone is de-ironed, we recheck their iron studies. If the patient is de-ironed, with a transferrin saturation of less than 40 per cent we wait three months and recheck. By then, the hemoglobin will usually be normal again at 150 to 175 for men and 120 -140 for women.

IF: It appears that the only test result you look at is hemoglobin. What about hematocrit? Do you check the transferrin saturation percentage and the ferritin levels at all during this phase of treatment and if so, how often?

Dr. Erb: During the de-ironing phase, I look at only the hemoglobin. Once they have been successfully de-ironed, I look at the percentage saturation, the hemoglobin and the MCV, which is the average size of the red blood cell. When they are perfectly stable, they should have a normal hemoglobin, a normal MCV and a normal saturation.

IF: Many patients report being very fatigued after each phlebotomy treatment, sometimes for days. In fact, it is the most common complaint we hear about and seems to cause the most disruption to their lives. What causes this?

Dr. Erb: I must confess, I don't typically have

patients who complain of that. If they do, it's usually only with the first. If they are fatigued after each phlebotomy, I expect it's related to the decreased blood volume. One could combat that by drawing off less blood, or alternatively, by giving them 500 ml of IV fluid at the time of their phlebotomy.

IF: Some people tell us that they have high saturations but a normal ferritin. What do you do with those individuals?

Dr. Erb: As I said before, in patients with hemochromatosis, the transferrin saturation always goes up before the ferritin. If they have hemochromatosis, they should undergo phlebotomy. Usually, I would do only six and then recheck their saturation. Unfortunately, although individuals with high saturations may not develop much liver disease, the increased iron can lead to heart disease. We see this particularly in type IV hemochromatosis patients who have normal saturations but very high ferritin levels. Despite the often dramatically high ferritins, they typically develop little if any organ damage. Phlebotomy treatment doesn't work well for them so chelation therapy is the alternative when required.

IF: We get a lot of questions from people asking about chelation therapy, especially from those who do not tolerate the phlebotomy therapy well. What is it?

Dr. Erb: Chelation refers to the removal of metals by chemicals that bind to them so that they can be excreted in urine. Desferrioxamine (Desferal) is the only iron-chelating agent approved for use in Canada. The gut does not absorb it so it must be given by other means. The treatment typically involves desferrioxamine 2-4 gms given by continuous subcutaneous

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Supplemental Reading

For more information on how excessive iron in specific tissues promotes infection, neoplasia, cardiomyopathy, arthropathy and endocrine/neurodegenerative disorders go to the Center for Disease Control's website www.cdc.gov/ncidod/EID/vol5no3/pdf/weinberg.pdf, and read Dr. Eugene Weinberg's paper "Iron Loading and Disease Surveillance."

While you're at the CDC website – www.cdc.gov/ – enter "hemochromatosis" in the search box for more in-depth information on different aspects of the disorder.

For the really keen, the physician training module is also well-done, and dovetails nicely with the information provided by Dr. Erb, although some differences are evident because of the different jurisdictions. www.cdc.gov/hemochromatosis/training/

Planned Giving Benefits the Giver and the Receiver

Many individuals consider planned giving as a way to be remembered, as a way of giving to their favourite charity and as a way of lessening taxes and probate fees. Financial planning can address the ongoing needs of a family, and provide a lasting contribution to the Canadian Hemochromatosis Society.

Many financial instruments exist that allow donors to take advantage of this giving method.

Buy Gas at Husky!

Every time you make a purchase at a Husky/Mohawk gas station, store or restaurant, and have your loyalty card swiped, CHS will receive 2 per cent of your purchase. A gas fill of \$45 means 90 cents is paid to CHS. Contact our office for your card today. Lottery and tobacco products, of course, are not eligible.

Donate Your HBC Reward Points

Zellers, The Bay, and Home Outfitters now issue HBC Rewards points. Help us by donating your points to the Society. Use our card #593 471 099 under the name CHS. Be sure to tell the rewards centre that you want to keep your own card active when donating points, or they will cancel it.

Good Donations

You can donate online through our website. Visit www.canadahelps.org. Search "hemo," then click "Donate now." This is a secure site. You can use your credit card with confidence.

Matching Gifts

Does your employer have a matching gift program? If so, please indicate the company name on your donation. If you aren't certain, just send us your employer's name and we can follow up. Many firms will match some portion of their employee's charitable donations.

When sending money . . .

. . . such as a cheque or Visa number, be sure to let us know what it is for. Money will be automatically entered as a donation unless you specifically tell us it is for a membership or in memory of a loved one.

- Bequests in a will
- A gift of property
- Life insurance
- Gifts of securities and stocks
- Endowment funds
- Charitable remainder trusts
- Charitable gift annuities
- Gifts of residual interest

Bequest In A Will

A lump sum or a percentage of an estate can be left to CHS. A receipt will be issued to offset estate taxes and taxes arising from capital gains.

Gift of Property

Any property can be donated through a will.

Life Insurance

An individual can purchase life insurance with CHS as beneficiary, with tax receipts issued for premiums paid. Current policies with cash value can also be donated and a tax receipt given for current value. Life insurance is not taxable when paid out as a death benefit and may be used to offset taxes owing.

Gifts of Securities and Stocks

Tax incentives that encourage a charitable gift of stocks and securities can be very attractive to donors.

Endowment Funds

Charities establish endowments to provide a source of future income. Donors may stipulate that a gift is be used for the charities endowment fund.

Charitable Remainder Trusts

Donors can transfer cash, securities, property, bonds etc to a trust. Income generated by the trust is then paid to the charity. When the trust ends or the donor dies, the remainder is distributed to the charity. The donor receives a receipt when the trust is established, based on the market value at the end of the trust. These assets do not form part of the donor's estate, and will reduce probate fees.

Charitable Gift Annuities

Annuities can generate a regular income for a donor. The charity is given a gift of capital, and buys a lifetime annuity for the donor. The income from this annuity is payable to the donor and is tax free.

Gifts of Residual Interest

An individual can deed a property to charity, and retain the use of the property for life, or other predetermined time, and receive a charitable receipt for the present market value of the residual interest. The advantage to the donor is reduced taxes and probate fees.

You can help CHS by making a planned gift to the Society. We recommend that you consult with your financial advisor, lawyer and life insurance agent. Call our office for more info.

Give Us Your Used Printer

If you have recently replaced your home or office printer with a better model, don't toss it before you check with us. The CHS office is looking for a donation of a small, working laser colour printer. Call our office and we will arrange shipping, or if you are in the Vancouver/Richmond area we will arrange for pickup. Thank you!

Of course a receipt will be issued for income tax purposes.

Donors, Sign in Please

When you go to the Canadian Blood Donor donor clinic on Oak St. in Vancouver, be sure to sign the life book at reception under "Canadian Hemochromatosis Society."

The Society is listed in the book, and we want to qualify to have CHS posted in the Life Link Board in the Donor Clinic.

Enjoy your newsletter!

Please pass it on. Our newsletter is also available online on our website. If you would rather read it electronically, or if you don't want future newsletters, let us know and we'll take you off the list.

Speak Up!

When leaving a message on our toll-free line, 1-877-BAD-IRON, leave your full name and address (spell them out) and your 10-digit number.

Notes from the International Bioiron Society Conference

The first congress of the International Bioiron Society was held in the Czech Republic at the Prague Hilton Hotel, May 22-26, 2005. It attracted approximately 500 participants from dozens of countries. The proceedings were dominated by numerous basic science “breakthroughs” in iron metabolism. The major theme of the conference was the iron-regulating peptide (short protein) Hepcidin.

Hepcidin is produced in the liver and its major function is to down-regulate the rate at which intestinal cells and macrophages release iron into the blood stream. Under normal conditions, the amount of iron transferred into the blood stream will be appropriate to body needs and excessive iron deposition in tissues will be avoided.

The gene which controls Hepcidin synthesis is called Hepcidin Anti-Microbial Peptide (HAMP). Hepcidin production goes up in

response to microbial infections and is one of the defences mounted by the body against invasion by microbes such as bacteria.

The hereditary hemochromatosis (HFE) gene modulates the expression of HAMP and this is probably the most important function of HFE. Hemojuvelin (HJV) – the “juvenile hemochromatosis” protein – also controls Hepcidin production, and the absence of Hemojuvelin leads to uncontrolled accumulation of iron in tissues.

Hepcidin appears to be the key to iron absorption and distribution within the body.

In discussions on hereditary hemochromatosis, the conference proceedings were bitter-sweet. While widespread population screening can no longer be justified, awareness of the disorder has increased enormously around the world. Also, a new iron chelating agent may have a role in selected individuals who are difficult to

phlebotomize. This new drug is Deferasirox (ICL670). It is a tablet which is generally well tolerated (in approximately 1,000 patients, mostly thalassemia sufferers, studied so far) with mild transient skin rashes and gastro-intestinal disturbances being the most common adverse effects.

The European Union apparently has funding available to support hemochromatosis-related endeavours in Europe. A brief meeting was held (chaired by Professor Pierre Brissot, Rennes France) to discuss co-operation between all the existing societies around the world. I attended the meeting on behalf of the Canadian Hemochromatosis Society and requested that we be considered for membership in this European club. There was broad support for the concept of membership from outside Europe. Minutes of this meeting will be sent to me in due course.

By Dr. Sam Krickler

Treatment of Iron Overload

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ous infusion overnight, 2-5 nights a week. It is much less efficient and less safe than phlebotomies and is therefore not indicated as standard therapy in types I to III hemochromatosis. It is only used as a last resort when phlebotomies are not well tolerated or occasionally in the case of acute arthritis caused when iron is initially mobilized by phlebotomies. This is treated with an intravenous infusion of 500 mg of desferrioxamine over two hours, once a week. A few doses are all that are required, then one can go back to phlebotomies.

Chelation therapy is also indicated for treatment of secondary iron overload caused by hemolytic anemia and bone marrow failure. In the first case, there is an increase in the breakdown of red blood cells so that they no longer live 110 days. This breakdown causes iron to be released into the bloodstream and accumulate in the tissues, just as in hemochromatosis. In bone marrow failure, the bone marrow does not make enough red blood cells. Patients are

therefore transfused, and again, become iron overloaded. Since both of these conditions involve an anemia, and the bone marrow is not capable of keeping up, phlebotomy is not an option.

IF: Is the kind of treatment regimen you have outlined here something a primary care physician could handle, and where would one go for phlebotomy therapy?

Dr. Erb: A primary care physician could certainly manage hemochromatosis, if he or she is aware of all of the problems that may present, such as diabetes and impotence, heart disease, other endocrine disease, and the need for vaccination. However, if a patient has significant disease, referral to the appropriate specialist would be advisable. Management of patients with significant damage to the liver is a whole other topic.

I will just say that at their initial meeting, patients should be screened for hepatitis A, B and C. If they don't have protective antibodies to hepatitis A or B, I give my patients a requisition to get vaccinated. Vaccination is provided free to hemochromatosis patients by

the BC government.

For the phlebotomies, out patient clinics at hospitals are an ideal place where the procedure can be done, and of course, if eligible, at regular blood donor clinics once the maintenance phase has been reached so that the 56 day interval is not a problem.

IF: The maintenance phase? So getting de-ironed isn't the end of phlebotomy treatments?

Dr. Erb: Just like height, weight and eye colour, the rate at which a person reaccumulates iron varies. After initial de-ironing, most individuals will need to have additional phlebotomies from two to six times annually to maintain transferrin saturation between 30 and 40 per cent. Saturations are preferable to ferritin levels as saturations always increase first. If their tests for hepatitis B and C were negative, and they meet all the additional requirements of the Canadian Blood Services, they can donate blood. If they cannot donate blood for any reason, they continue their phlebotomies where they've been having them done. Treatment is ongoing for life.

Be A Movie Star!

A documentary film is in development and the filmmakers want to talk to people with HHC who are also still very active and involved with other challenges in their life, while managing their HHC.

If you are running, swimming, playing tennis or any sport, or participating in a musical event or studying for a degree or writing a screenplay or anything that taxes you physically and mentally, you might be the perfect subject. Contact our office or the program producer directly at d-davey@sympatico.ca.

Contact us!

Canada Post #272 - 7000 Minoru Boulevard
Richmond, BC Canada V6Y 3Z5

Telephone 604-279-7135

Fax 604-279-7138

E-mail office@cdnhemochromatosis.ca

Toll Free 1-877-BAD-IRON

www.cdnhemochromatosis.ca

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Please call, write or email our office with your order. Our supply is limited, so don't delay. Send a cheque or process your Visa transaction easily and securely on www.canadahelps.org and specify in the notes that you are purchasing a band.

Be sure to indicate the number and size (small or large). Total cost for each wristband is \$7.50, which includes taxes and mailing. **DON'T FORGET TO INCLUDE YOUR MAILING ADDRESS!** We will send your band or bands by return mail. Wear them proudly!

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I have HHC A blood relative has/had HHC

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- Do not publish my name in any CHS media.

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 #272 - 7000 Minoru Boulevard
 Richmond, BC Canada V6Y 3Z5
THANK YOU!

October, 2005