



Welcome to the Winter Newsletter 2009



In this issue:

- Mick McGowan back in action
- Your Heart and Haemochromatosis
- Noel's Story

FEATURES

- Report from National Ploughing Championship
- Presentation by Dr Gerard Crotty in Tullamore
- Clare / Limerick Meeting in Shannon

MICK MCGOWAN BACK IN ACTION WITH HIGH HOPES

Mick McGowan (36) grew up in Duleek, Co Meath. He became Irish Senior Darts champion at 17 and at the age of 18 was the youngest captain of the Irish team. Having recovered from a serious car accident which threatened to end his career, he began competing in Professional Darts Corporation (PDC) tournaments in 2004.

In 2006 he entered the full PDC circuit, winning two ranking competitions, The Irish Open and The Vauxhall Classic. He hit 25 in the world rankings early in 2007. Then things began to slide a little.

He was extremely tired and experiencing severe joint pain that just did not seem to go away. He attributed his chronic fatigue to his heavy schedule of competitions and travel in Europe. Eventually in 2008 his wife suggested he consult his GP.



At this point with a ferritin level of 2,500mg, Mick was diagnosed as having Haemochromatosis. Mick acknowledges that his condition obviously contributed to his poorer performance at darts. Prior to his diagnosis, his continuous battle with pain and fatigue meant that he had found it increasingly difficult to put in the practice hours needed to keep on top of his game.

This year Mick is feeling much better as his iron levels have been brought down to normal by the venesection treatment. His energy is greatly improved and he is now able to practise three hours a day.

We wish Mick every success in the future and thank him for sharing his story which has been adapted from an article in the Irish Times Sports Section by Sean Kenny, September 19th 2009.

NATIONAL PLOUGHING CHAMPIONSHIP (NPC)

Once again the IHA took a stand at the National Ploughing Championships in Athy Co. Kildare. We were blessed with perfect weather and enjoyed three great days. We would like to thank the many members and friends who helped to man the stand and administer the survey that we conducted.

Approximately 180,000 people attended the NPC and we met a wonderful cross section of Irish society. This was an ideal opportunity for us to gain a better understanding of "who knew what" about Haemochromatosis.

- A total of 647 people responded to our survey and a snap shot of the results are shown below.
- 61% (395) had heard of Haemochromatosis
- 41% (263) of those who had heard of



Seamus Leydon (GAA All Star!) and Rose Tubridy helping out at the NPC

- HH, knew someone who has it
- 5% approx (31) have HH
- 30% of those with HH are currently undergoing treatment
- 74% of those who need to give blood would be willing to donate to the



Philippa Moran, Tony Alyward and Vivienne Clarke who also volunteered for the day

Irish Blood Transfusion Service at an IBTS clinic convenient to them or at an IBTS Blood Mobile.

The IHA continues to campaign for the nationwide extension of the Stillorgan pilot programme.

YOUR HEART AND HAEMOCHROMATOIS

Arrhythmia with shortness of breath and chronic fatigue might be a sign of cardiomyopathy. Cardiomyopathy is a disease of the heart muscle where the muscles may be stretched (dilated), enlarged (hypertrophic) or cannot pump properly (restrictive). Restrictive cardiomyopathy is sometimes found in those who have Haemochromatosis. Transferrin, a protein, binds to iron we absorb and moves it to storage in ferritin or to bone marrow cells where haemoglobin can be made. When transferrin is highly saturated and can no longer bind to iron, excess iron may become get free and become a dangerous oxidant or begin to precipitate as haemosiderin. Ferritin contains mobilizable iron, the type that can be removed with venesection treatment. Haemosiderin is not mobilizable; it cannot be moved out of the cells. Instead, it accumulates in tissues damaging the

targeted organ, impairing its ability to function. Ferritin levels tell the physician approximately how much iron has accumulated in various organs. The heart is especially vulnerable because it produces less ferritin than other organs, like the liver, spleen and bone marrow. Therefore membranes of the heart are prime targets for iron to accumulate, impeding function of cardiac muscles to pump blood efficiently and possibly lead to a heart attack. Transferrin iron saturation percentage reveals just how saturated the transferrin is with iron. When the percentage is high, transferrin is not going to be able to efficiently bind to iron. Therefore any excess iron that a patient absorbs that cannot be bound to transferrin will become free. This can nourish cancer cells or load in the heart and other organs as haemosiderin.

Venesection involves removing one unit of blood per week for a period of time, while observing haemoglobin. In an iron overloaded patient, haemoglobin will rebound after blood extraction. Venesection treatment should continue until ferritin is within a safe range of 25-75 ng/ml. Then a patient will follow a maintenance program as recommended by the attending physician. Maintenance may involve as few as 2 to 3 blood donations per year, and simple changes to diet and exercise. Cardiac Diagnostic Aids such as Electrocardiogram (ECG) or Magnetic Resonance Imaging (MRI) will not usually reveal iron in the heart.

This article was adapted from the Iron Disorders Institute newsletter, 2nd Quarter 1999 and was written by David Meyers, MD. University of Kansas Medical Center.

NOEL'S STORY

Diagnosis

'My diagnosis of Haemochromatosis was prompted through the investigation of blinding headaches that I was suffering from and which were not being brought under control by the use of the painkillers prescribed to me by my GP. In 2001, having just turned 50 and taken up the position of General Manager of a busy hotel in County Meath, I began to experience very severe headaches. They did not respond to the painkillers prescribed to me by my GP and in 2003, I was referred to a Neurologist in St James's Hospital Dublin. As part of the process a series of blood tests were carried out and the result of one test caused concern. My ferritin level was 1,900ng/ml and I was referred to a Haematologist in St James who diagnosed Haemochromatosis and referred me to Prof. Suzanne Norris who takes expert care of my condition.

Treatment

For the year and a half following my diagnosis, Nurse Liz Ellis took a unit of blood each week in order to bring the iron down to the level of 50 ng/ml. It suited me to attend for venesection every Monday morning at 8.00 and to return to work afterwards. However, I will never forget how awful I felt

approximately two hours after each venesection, at one time feeling so bad that I thought I would collapse. It seemed that I could not tolerate giving blood on this regular basis. Here Prof Norris was again of great help to me as in consultation with the Haematologist she decided that I should have an injection of Epo 24 hours in advance of the venesection which would increase my red blood cells. Thankfully, this was successful and I was shown how to administer the Epo injection myself.

One other complication for me regarding the venesection was the fact that my veins are very deep. It was therefore necessary for Nurse Liz to contact a doctor in Haematology to find a vein prior to each venesection.

The results of a liver biopsy thankfully confirmed that I did not have liver damage.

I continue to attend St James's under the care of Prof Norris. Every six months she checks my blood levels and organises an ultra sound of my liver and other organs to make sure that there are no signs of disease.

Life before and after my diagnosis

Before my diagnosis of HH, I remember playing a round of golf with a friend from the U.S. After the fourteenth hole, I became so

exhausted that I had to call it a day. Soon afterwards I found that even nine holes was too much of a challenge for me and I allowed my golf club membership to lapse. Since my diagnosis and now that my ferritin level has been brought under control, my energy level is as good as it was in my twenties. I rejoined the golf club and can play a full round of eighteen holes without any difficulty.

And the headaches

The headaches were not related to my Haemochromatosis and I had quite a long journey towards a cure - attending my GP, three neurologists and a faith healer en route. Finally, in 2007, I was referred to the Endorphin Release Clinic in Drimnagh and to my great relief, the cause of the headaches was diagnosed and treated. After seven years of severe blinding headaches, I no longer suffer from them.

I am very grateful to all the health care professionals in St James's Hospital who worked with me to manage my haemochromatosis and to point me in the direction of a cure for my headaches. Today, I enjoy a healthy, active life!'

The IHA would like to thank Noel and hope that his interesting account will help others.

DR GERARD CROTTY'S PRESENTATION in TULLAMORE on OCT 19th



Dr Gerard Crotty and Nurse Frances Neville

Dr Gerard Crotty, MRCPI is a Consultant Haematologist at the Midland Regional Hospital. He gave a comprehensive overview of the Diagnosis and Treatment of Haemochromatosis. Dr Crotty's presentation was complemented by a presentation from Nurse Frances Neville who is a Specialist Haematology Nurse at the Hospital. Nurse Neville gave us an insight into the nurse's perspective in dealing with HH. The account below is extracted from Dr Crotty's presentation.

What is haemochromatosis?

- Iron overload disorder
- One of the commonest genetic disorders (in European populations, especially Irish)
- Treatable with a simple treatment which can reverse most of the potential problems if diagnosed in time
- Not curable – needs ongoing maintenance treatment

What are the symptoms?

- May be none
- Tiredness
- Joint pains
- Loss of sex drive
- Liver disease
- Heart failure (cardiomyopathy)/ Irregular heartbeat
- Diabetes

What causes HH?

- Gene – called HFE
- 2 mutations – C282Y and H63D

Commonest way to have the disease

- C282Y / C282Y (homozygous)
- Some patients have C282Y/ H63D
- H63D / H63D

Other genetic findings which do not cause disease

- C282Y / Normal
- H63D / Normal

How do we diagnose it?

- Suspected from symptoms
- Family history
- Screening test

- Ferritin raised implies increased Iron stores but nonspecific
- Transferrin saturation more specific but test is subject to influence of diet/time etc
- Genetic test

Iron in the body

- Normal adult – 3g iron
- No system for getting rid of excess iron
- Loss of 1mg/day by loss of cells
- Women – loss in periods / pregnancy
- Normal diet – 10mg/day
- Only 1-2mg absorbed
- If too much iron gut cells block this
- Block doesn't work in HH

Loss of sex drive

- May be loss of desire
- Or loss of performance – inability /difficulty in getting or maintaining an erection (Erectile dysfunction)
- Treatment : Exclude other causes – diabetes, blood pressure, medications
- Testosterone replacement – gel
- Tests for organ damage
- Liver blood tests
- Glucose (for diabetes)
- Alpha –fetoprotein
- Ultrasound of liver
- Echocardiogram
- Hormonal tests eg testosterone
- Liver biopsy

Liver biopsy

- Before a genetic test was available the liver biopsy was often needed to make the diagnosis
- Now needed in some patients to diagnose cirrhosis of the liver (a complication)
- Only needed in those more severely affected

What happens after diagnosis?

Venesections

- Every week until target
- Can be every 2 weeks in mildly affected patients
- Then every 3 months (varies) to keep on target
- Treatment for any complications

Target values

- Ferritin < 50 (Normal is 10-320!)
- Maintenance venesections every 3 months initially
- Other doctors use variations on this
- Some recommend keeping transferrin saturation < various values
- Some let the ferritin rise and do a number of venesections to bring it down

Diet

- Avoid iron supplements
- Avoid Vitamin C supplements >500mg/day
- Moderate alcohol intake (or stop completely if liver disease)
- In treated patients, we don't recommend other dietary changes
- May be helpful to reduce meat etc in patients with borderline iron overload

What about family members?

- Siblings (brothers and sisters) should be tested using the genetic test
- Children – only at risk if other parent also has mutation – test parent
- Not advised to test younger children

Inheritance

- Recessive : To have the disease, you must get an abnormal gene from both parents
- People with only one abnormal gene are called carriers
- If both parents are carriers there is a 25% chance that the child will have the two genes.



CLARE/LIMERICK MEETING - MONDAY 9th NOVEMBER 2009



Dr Denis O'Keeffe who spoke at the meeting in Shannon

The IHA would like to express sincere thanks to Denise McAuliffe who organised the meeting which took place in the Oakwood Arms Hotel, Shannon, Co Clare.

Our thanks also to Dr Denis O'Keeffe, MRCPI, Consultant Haematologist at the Midwestern Hospital, Dooradoyle, Limerick, and Sr Fiona Joy, Clinical Nurse Specialist in Haematology, who both gave excellent presentations on the night.

A very interesting question and answer session followed the talks and addressed relevant issues of concern to HH patients. One such issue relates to sexual dysfunction and Dr O' Keeffe suggested that patients who experienced problems should consult an endocrinologist. On the

subject of alcohol consumption by HH sufferers Dr O'Keeffe stressed the importance of keeping alcohol consumption within the recommended limits as alcohol can increase iron absorption.

Issues of concern are outlined briefly below:

- Anger and frustration at the fact that the Limerick Regional Hospital is no longer accepting new patients for venesection. This makes it very difficult, if not impossible, for some patients to access venesection.
- Another serious problem is the fact that there is no protocol in place for the disposal of venesected blood by GPs
- The difficulty that GPs experience in accessing venesection kits. It is hard to accept that there is not a consistent national approach to the problem that would enable GPs to provide this service if they choose to do so.
- VHI policy towards venesection. A 25 year old man who attended the meeting was told by the VHI that he would not be covered for venesection until he was a member for five years! It is very hard to understand the rationale behind this decision.

ADDITIONAL INFORMATION

THANKS TO BRENDAN GALLAGHER

We would like to thank our outgoing treasurer, Brendan Gallagher, for the great work he did on behalf of the Association over the last few years. His help has been invaluable and is very much appreciated.

Brendan Keenan who is a Director of the IHA, has taken over this very responsible and time-consuming task. Brendans address is: 20 Templemore Avenue, Rathgar, Dublin 6

MINI-MARATHON

Thanks to all members and friends who helped to raise a record €6,880 for the IHA. Sinead Kissane, a well known sports presenter with TV3, ran the marathon again this year on behalf of the IHA and donated the €1,000 sponsorship given to her by Flora to the Association.

RENEWAL OF SUBSCRIPTION

Membership renewal forms are enclosed with this newsletter. Please complete and return to Brendan Keenan. We would like to remind our members that your ongoing support through the annual subscription is very much appreciated and is one of the main sources of income for the Association.

If you no longer wish to be included in our mailing list, we would appreciate it if you would let us know as the cost of posting and printing is high. If your email address has changed or if you would prefer to receive the newsletter by email please let us know.

The IHA would like to wish everyone a Happy Christmas and a peaceful New Year.

Disclaimer: The IHA believes the information in the newsletter is accurate, but little is known about many aspects of HH and research is progressively revealing new information on the subject. Accordingly, any person using this newsletter does so, on the condition that he or she thereby indemnifies and keeps indemnified the IHA against action or any claim of any nature whatsoever arising directly or indirectly from the use of information contained herein.

OTHER REGIONAL MEETINGS WHICH HAVE TAKEN PLACE

SLIGO

Date: Thursday November 19th 2009 at 8pm
Venue: Clarion Hotel, Clarion Road, Sligo
Speaker: Dr Kevin Walsh M.D MRCPI, Consultant Gastroenterologist Sligo General Hospital



Margaret Mullett and Dr Kevin Walsh at the Sligo Meeting

CORK

Date: Tuesday 24th November 2009 at 7:30pm
Venue: Maryborough House Hotel, Douglas
Speakers: Dr Orla Crosbie - Consultant Gastroenterologist
 Dr Mary Cahill - Consultant Haematologist Cork University Hospital
 Dr Ann Nicholson - GP, Ballincollig, Cork

Despite severe flooding and a change of venue, there was a record attendance of over 90 people, some travelling from as far as Castletownbere and Limerick.



Dr Orla Crosbie, Dr Mary Cahill and Dr Ann Nicholson at the Cork meeting

KILKENNY

Date: Tuesday December 1st 2009 at 8pm
Venue: Newpark Hotel, Castlecomer Road, Kilkenny
Speaker: Dr Brian Hennessy M.R.C.P.I Consultant Haematologist, St Luke's General Hospital Kilkenny & Waterford Regional Hospital