



## Welcome to the Spring Newsletter

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### ANNUAL GENERAL MEETING SATURDAY MAY 25<sup>TH</sup> 2013

The meeting will take place at the Irish Blood Transfusion Service (IBTS), St James's Hospital, Dublin 8 on Saturday May 25<sup>th</sup> 2013. The IHA would like to thank the IBTS for once again making the centre available to us and for generously sponsoring coffee and lunch. Coffee will be available from 10.30 am. A brief business meeting will commence at 11.15am and will be followed by guest speakers, Dr John Keohane, Consultant Gastroenterologist, Louth County Hospital and Mr Ray McKiernan who will speak from a patient's perspective. Dr Willie Murphy will give an update on the progress that the IBTS has made with regard to providing venesection for Haemochromatosis patients.

The talks will be followed by a Question and Answer session and will conclude with lunch. For catering purposes, please let us know as soon as possible if you are attending by returning the reply slip posted out with the official AGM announcement or by phoning (01) 8735911.

The address of the secretary is:  
Ann Campbell,  
7 Ashleigh Green,  
Castleknock  
Dublin 15

## LOUTH INFORMATION MEETING



The meeting at the Fairways Hotel, Dublin Road, Dundalk on Tuesday March 5<sup>th</sup> was one of the most successful meetings ever. Nurse Anna Cappalis, Nurse Majella Jobling and their colleagues organised the meeting. Approximately 250 people attended, including people with the condition, family members and practice nurses from the surrounding area.

The excellent speakers were Dr Sengupta, Dr John Keohane and Nurse Majella Jobling. The meeting was chaired by Nurse Anna Cappalis. The aim of this event was to help raise awareness of Haemochromatosis, including how to identify it and how to live with it.

The Haemochromatosis/Venesection clinic is based at the Louth County Hospital and covers the Louth/Meath region. Louth County Hospital already has 668 patients attending the clinic. Dr John Keohane, Consultant Gastroenterologist explained the signs and symptoms, genetic screening and management of the condition. He said that treatment for the condition is simple and effective.

Dr Sengupta, Consultant Gastroenterologist is the lead clinician and provides governance for the clinic. He spoke about the referral model developed in the Louth/Meath region. Patients start their treatment programme (venesection) within three weeks from the date of referral. This



From left: Dr John Keohane, Dr Sengupta, Nurse Anna Cappalis and Nurse Majella Jobling.

model is working very well and it was suggested that this system could be used on a national basis as it would prevent patients having to wait long periods of time to see consultants prior to commencing treatment. Dr Sengupta also spoke about the need for a national database which would help standardise record keeping and improve research into the condition. The Irish Haemochromatosis Association sponsored the event and the treasurer Brendan Keenan travelled from Dublin to represent the association.

Margaret Mullet spoke about the work of the IHA and the importance of raising awareness. She asked for volunteers for the Awareness Day on June 6<sup>th</sup> and for the Mini Marathon on June 3<sup>rd</sup>.

A lively Question and Answer session followed the meeting and many queries were answered.



The Lord Mayor, Naoise O Muiri, hosted a reception at the Mansion House on Dec 7th for Brendan Kaye and Ben Shuker.

## HOW CAN REMOVAL OF BLOOD REMOVE STORED IRON?

Your body contains about 5 litres of blood. Your bone marrow is the blood cell factory of your body. Routinely in the human body, red blood cells are recycled every three months. If you lose blood by bleeding or donating blood, your bone marrow can replace that blood as long as you have enough iron, vitamin B12 and folate. This is because iron, vitamin B12 and folate are ingredients used to make new red blood cells.

After blood is removed by a venesection, some of the iron that is stored in the body moves out of storage and becomes available to help make new red blood cells.

If you have Haemochromatosis and are iron-overloaded and have adequate vitamin B12 and folate available, your bone marrow will be able to make new red blood cells every time you have venesection

A standard 500ml venesection removes 0.25 grams of iron from the body. (That is equivalent to the amount of iron in a 50mm nail). It takes four venesections to remove 1 gram of excess iron (four nails).

A person with moderate iron overload may have between four and 10 grams of excess iron, which will take between 16 and 40 venesections to reduce to normal

levels.

Severe iron overload may be up to 40 grams of excess iron which will require up to 160 venesections to reduce to normal levels.

### BLOOD TESTS

If you have ever had a blood test that shows you have increased iron levels or that you have abnormal liver function then you should be tested for Haemochromatosis.

If both the Serum Ferritin and Transferrin Saturation (T.S.) are raised then the genetic test should be done. The abnormal Ferritin range for females is above 200ug/l and for men above 300ug/l. Regarding T.S., the abnormal range for females is above 45% and for men above 50%.

There are a number of reasons apart from Haemochromatosis that can lead to increased iron levels and abnormal liver function. These include Hepatitis B infection, Hepatitis C infection, alcoholic liver disease and fatty liver (also called Non-Alcoholic Fatty Liver Disease).

*(This Information was provided by Professor Lawrie Powell and Prof John Olynyk for Haemochromatosis Australia)*

## GUTHRIE PIN-PRICK CARDS

The Irish Haemochromatosis Association is a member of the coalition group who are lobbying the Minister for Health to ensure that the valuable archive of Guthrie cards held in Temple Street Hospital is not destroyed.

Since the Irish National Screening was established in 1996 more than 2.5 million samples have been put on Guthrie cards, analysed and stored.

Thousands of children have been protected from the consequences of PKU and other genetic disorders.

At present six disorders have been tested for including Cystic Fibrosis and Phenylketonuria (PKU).

The Guthrie cards were used by a research team from the Mater Hospital to establish the fact that one in 83 Irish people are predisposed to iron overload.

This valuable information has alerted doctors to the importance of screening for Haemochromatosis.

**Good News:** The latest update is that as a result of the work of the coalition group, Minister Reilly has asked for an expert group review instead of going ahead with the destruction of the samples. The Minister will be looking for information as to how other European states avoided destruction of the cards while staying within the remit of European data protection law.

## COFFEE MORNING

A very successful coffee morning was held in Bushy Park Rd, Dublin on Thursday March 14<sup>th</sup>. Over €1000 was collected and a very sociable event was enjoyed by neighbours and friends.

Haemochromatosis brochures and an invitation to the coffee morning were circulated locally and it proved a great way of raising both money and awareness. If anyone would like to organise a similar event, please get in touch and we will send on brochures.

## THE ROLE OF HEPCIDIN IN REGULATING IRON ABSORPTION

Hepcidin has now been shown to play a critical role in the regulation of iron absorption from the diet. It has also been shown that patients with all forms of HH have low or undetectable levels of this molecule, leading to inappropriately increased absorption of iron from the diet relative to the level of iron stores already in the body.

In contrast to this, patients with anaemia of chronic inflammation (also known as anaemia of chronic disease) have too much Hepcidin, resulting in not enough iron being released into the blood stream.

In the future, HH patients may be treated with Hepcidin injections in order to control

the amount of iron taken up from the diet, similar to the manner in which insulin injections control the amount of sugar in the body.

For patients with anaemia associated with too much Hepcidin, the development of drugs to block Hepcidin interacting with ferroportin may lead to the release of more iron into the blood stream. The use of novel intervention systems to inhibit or increase Hepcidin function will require the availability of a reliable robust assay system for the measurement of Hepcidin levels. Scientific researchers interested in Chronic Liver Disease (CLD) have looked at expression of Hepcidin in HH livers and

demonstrated that it is indeed lower in HH than in normal livers. The next step is to measure serum Hepcidin to ascertain the levels of this protein in those that are not yet loading iron versus those who are iron-loaded. They will also examine the effect that phlebotomy has on Hepcidin levels and will correlate these levels with other biochemical iron indices in both HH and in those individuals who do not have C282Y or H63D. Hopefully then it will be possible to work out firstly what exactly signals the liver to produce Hepcidin and secondly why HH patients do not produce enough Hepcidin despite being iron loaded.



# IRON OVERLOAD

## BY TOM MULVIHILL

I was born in Ballydonoghue, Lisselton, Co Kerry. I enjoyed a healthy and active life.

I joined An Garda Siochana in 1974. My first posting was to Monaghan Town. I came to Limerick in 1978 and retired from Bruff in 2004. During my service I had worked a three relief shift system. I found the system to be very severe on my health with broken sleep patterns and irregular meal times. I noticed that for over thirty years, I was very prone to getting sore throats which invariably lead to chest infections which required antibiotics.

From about 1990 on I noticed that I would get very tired for no apparent reason while driving and would have to pull in and get 40 winks even on a half hour journey. I presumed that the tiredness was caused by irregular working hours. In late 2003 I really felt unwell and had lost about a stone in weight. My pale colour was very noticeable and a number of people had commented on my appearance. If I had any alcohol I would be unwell for two days.

In January 2004 I was referred to Dr Manus Moloney, a Gastroenterologist based at Nenagh General Hospital and the Mid-Western Regional Hospital. After a clinical examination he considered that I was suffering from iron overload and a blood test revealed that I had ferritin reading of 4,400 when the reading should be fewer than 300mg. A genetic test later confirmed that I had Haemochromatosis. On being informed of this condition I didn't know whether to laugh or to cry so I did a bit of both. At

last I had a diagnosis of my ailment which was a relief. However, it turned out that I had developed severe liver damage due to the iron absorption. Due to the high level of iron he decided that I would require urgent and intensive treatment. In all I had 150 pints of blood drawn off in two years and three months. I would need to rest for some two to three hours before returning home after a venesection whereas some of my fellow patients would be gone out the door in a matter of minutes. It would take me two or three days to recover.

Dr Moloney had not come across another patient with a ferritin as high as 4,400 and he has used my case history as part of his presentations on the subject. Haemochromatosis is more of a life sentence than a death sentence. The treatment is relatively straightforward-bloodletting on a regular basis.

As I would be reduced to working light duties I decided to take early retirement to look after my health and my family. This decision came easy and I have no regrets. I am now on what is called maintenance stage which involves removing about eight pints of blood annually. I try to have a regular diet but must avoid any iron supplements and alcohol. My overall health has improved as has my colour. I rarely get chest infections due to my improved immune system. Life is good and I am lucky to have been diagnosed before further damage was done.

*(The IHA would like to thank Ex-Sergt. Tom Mulvihill for this interesting account and for his on-going help to the IHA).*

### UPDATE FROM DR PATRICK KEILY ON THE HH ARTHROPATHY CLINIC, ST GEORGE'S HOSPITAL, LONDON

'Seventeen patients have been sent so far to the clinic which is held every alternate Monday afternoon, with referrals coming in from willing GPs as far afield as Shropshire and Yorkshire. We have had two samples of joint tissue sent in by willing surgeons. It is fascinating and most instructive to hear the stories and tribulations of patients, particularly how the diagnosis was made in the first place, when the joints became involved, and what has or has not been helpful. I am very grateful to those of you who are prepared to give your time to travel to St George's to let me record

your details, examine your joints, arrange scans and add your data to a growing database of information about the arthropathy of HH. As my accumulated experience grows, so I hope to get a clearer understanding of the arthritis and share with you what we might be able to do to ameliorate or stop it in the future. It would be a pleasure to meet as many patients as possible, so if you are able to travel to St George's please try and get a referral from your GP.

Patrick.Kiely@stgeorges.nhs.uk 0208 672 1255

*(Thanks to the British Society for allowing us to include this interesting information)*

### LIVING WITH HAEMOCHROMATOSIS BY A MEMBER OF THE IHA

The outlook for people who have Haemochromatosis largely depends on how much organ damage has already occurred at the time of diagnosis. Early diagnosis and treatment of the disorder are important. Treatment may help prevent, delay, or sometimes reverse complications of the disorder. Treatment also may lead to higher energy levels and better quality of life. With early diagnosis and treatment, a normal lifespan is possible. If organ damage has already occurred, treatment may prevent further damage and improve life expectancy. However, treatment may not be able to reverse existing damage. If Haemochromatosis is not treated, it can lead to severe organ damage or even death. All of the above will be familiar to members of the Association. However, there is one feature of Haemochromatosis that is seldom mentioned but which should be faced. This is the effect that an accumulation of iron in the pituitary gland has on various hormonal functions. Accumulation of iron in the pituitary gland can interfere with the pituitary's control of sex hormones. In men, pituitary damage can lead to impotence, decrease in testicular size and even osteoporosis (bone weakening) leading to fractures. About 45 percent of men who have symptoms at the time of diagnosis have impotence. Fortunately, loss of libido (sex drive) often accompanies impotence, thereby lessening the significance of the latter. In women, pituitary damage can lead to amenorrhea (absence of menstrual periods), although women with Haemochromatosis seldom experience loss of libido or premature menopause. Various treatments for impotence have been promoted, including the use of mechanical devices as well as medication. The inconvenience of the former, in addition to its effectiveness, is a deterrent to its use. The appropriateness and effectiveness of medication appears to be doubtful.

**FLORA WOMEN'S  
MINI MARATHON,  
DUBLIN, BANK  
HOLIDAY MONDAY,  
JUNE 3rd 2013**



We urge all members to persuade friends, family and colleagues to run, jog or walk the 10K route for a great day out. Sponsorship cards and T shirts are available from Margaret Mullett on 01 4922705 or email: margarettmullett@gmail.com  
Entry Closing Date: 24th April 2013 OR when maximum number of entries is reached.

## HAEMOCHROMATOSIS AWARENESS DAY, THURSDAY JUNE 6<sup>TH</sup>

There will be information stands in several shopping centres throughout the country. If you can assist for even a few hours please contact: margaretmullett@gmail.com

It would be appreciated if anyone with contacts in the media would get in touch with us. We need to highlight this Awareness Day in the papers, on radio and TV. Our aim is to help save lives by alerting people to the possibility that they may be suffering from Iron Overload. Your help is essential to the success of this day. Please phone 018735911 if you can help in any way or email: margaretmullett@gmail.com

### PROPOSED VENUES

The following venues have given permission for us to have information stands. This will only be possible with the help of volunteers to man the stands.

In Dublin there will be stands in:

1. St Stephen's Green Shopping Centre
2. The Ilac Centre, Henry St
3. Nutgrove Shopping Centre, Rathfarnham
4. Swan Centre, Rathmines
5. Ashleaf Shopping Centre, Crumlin
6. Frascati Shopping Centre, Blackrock
7. Blanchardstown Shopping Centre
8. Donaghmede Shopping Centre
9. Swords Pavilion Shopping Centre
10. Dundrum Shopping Centre

#### Cork:

11. Mahon Shopping Centre
12. Douglas Shopping Centre
13. Wilton Shopping Centre

#### Galway:

14. Corrib Shopping Centre
15. University College Hospital, Galway
16. **Waterford:** City Square Shopping Centre

17. **Wexford:** Gorey Shopping Centre
18. **Roscommon:** Molloy's Pharmacy

#### Limerick:

19. Crescent Shopping Centre
20. Charleville Shopping Centre
21. **Kilkenny:** Market Cross Shopping Centre
22. **Clare:** -Ennis- Dunne's Shopping Centre

#### Louth

23. **Dundalk:** The Marshes Shopping Centre
24. Louth County Hospital
25. **Sligo:** The Tesco Arcade Shopping Centre

26. **Mayo:** Castlebar Hospital

27. Meath Our Lady's Hospital Navan Hospital

#### Westmeath:

28. **Mullingar:** Harbour Place Shopping Centre
29. Athlone Town Centre Shopping Centre
30. **Kerry:** Tralee, Manor West Shopping Centre
31. **Kildare:** Newbridge Whitewater Shopping Centre
32. **Monaghan:** Monaghan Shopping centre

## ATTITUDE OF THE ARMY TO HAEMOCHROMATOSIS

The IHA recently contacted Minister Alan Shatter on behalf of young men who were disqualified from applying to the army as cadets because of a diagnosis of Haemochromatosis (HH). More recently the IHA was sent information by a serving member of the defence force who felt that soldiers were treated differently from officers.

### Minister's Reply:

'The professional advice of the Medical Military Authority is that haemochromatosis is a hereditary condition, which if detected in time, and with the implementation of required interventions, can lead to a full and normal life experience. However, if ignored, either intentionally or inadvertently, or if undiscovered, it can give rise to serious complications which can be life threatening. The position is that there are a number of physical and medical standards laid down by the Military Authorities for entry to the Defence Forces. These standards are applicable to all those applying, either as recruits or cadets.

If any individual is admitted knowingly to the Defence Forces with the genetic predisposition to haemochromatosis,

the Medical Corps professional advice is that a duty of care arises to monitor the development of the condition as well as to intervene with the necessary treatments in order to avoid the development of complications. Ultimately, there may be a requirement to actually treat the complications. Due to the requirement to carry out regular blood testing in order to detect or monitor progression, or potential progression, the candidate falls into a category outside the appropriate required standard for service in the Defence Forces. Medical standards, which must be achieved, for Extension of Service, Re-Engagement and Continuance in Service, are also set out in Defence Force Regulations. The maintenance of a cadre of personnel capable of meeting the physical demands of a career in the Defence Forces is a key human resources requirement. The medical classification of all personnel is kept under review and members of the Defence Forces are medically classified in accordance with Defence Force Regulations A12 and associated guidance Instructions. The Regulations and Instructions are framed in the context of the unique and demanding nature of the physical requirements of operational military life.

## SLIMLINE MILK FORTIFIED WITH IRON

Several members of the IHA have contacted the association with regard to the fortification with iron of Avonmore Slimline Milk.

The IHA contacted Prof Mark Worwood, who is an expert on iron overload, for his advice. He is one of the scientists who sat on an independent review on 'Iron and Health' conducted by the UK Scientific Advisory Committee in 2010. In his reply, Prof Worwood suggested

that 'Avonmore should either show that their product is not enhancing iron absorption in subjects with Haemochromatosis (in which case it does not work!) or should add a warning note advising subject with Haemochromatosis not to drink the product'. The IHA have been in touch with Avonmore who are very reluctant to take the expert advice given by Prof Worwood.

## NOTICE TO MEMBERS

We would like to thank all members who renewed their membership and those of you who so generously gave an extra donation. If you have not yet had the opportunity to renew, please do so as soon as possible. I am sure you appreciate that the cost of posting and printing has gone up considerably and the Association depends on your renewals to continue in its aim to raise awareness of this condition.

If you wish to receive the newsletter by email please let us know by emailing kategeog@gmail.com or leave a clear message on the answering machine

018735911. If you have a new mobile number or email address please send it to Kate as texting or emailing is the most economical and quickest way for us to contact you. We have continued over the years to send out information to everyone on the data base even when they have overlooked the renewal notice. Unfortunately we can not afford to continue doing this. Please let us know if any member would like to have their name deleted from the data base. **It is hoped to arrange an information meeting in Galway. Details will be on the website or phone 01 8735911**

**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.