Iron Disorders Institute receives standing applause from attendees—
Western Regional Hemochromatosis Conference—April 3-4, 2009: Reno, Nevada

“This year’s regional conference was an enormous success mostly because of the change to the format.” Comments Cheryl Garrison, Executive Director, Iron Disorders Institute (IDI). The format included a one half day CME activity for healthcare professionals held on Friday afternoon April 3rd and followed by an all day Saturday session for patients. The presenters were the same for both days except for Dr. Suchi Pandey, Assistant Medical Director, Blood Centers of the Pacific, San Francisco, who presented on Saturday only.

John A. McDonald, MD, PhD: Vice President, Division of Health Sciences University of Nevada, Reno kicked off the conference. Dr. Lewis Wessellius, IDI Medical & Scientific Advisory Board Member served as Program Chair. IDI Founding Director Chris Kieffer assisted Conference Events Director Mardi Brick. Mardi is pictured here relaxing after receiv-

John McDonald, MD, Ph.D.

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If we have your email address, we will send you a reminder that nanograms is available. Printed copies will be mailed to IDI MEMBERS. Join today and get your printed copy of nanograms.

COOK! For Iron-Balance Page 10

As I see it every day you do one of two things: build health or produce disease in yourself.”
— Adelle Davis

Start your morning with EGG Power ....page 10
Iron Disorders Institute Board of Directors Vice Chair Gerry Koenig recently attended the 2009 International Bio-Iron Society Meeting held in Porto, Portugal. Bio-Iron is held every two years and attracts hundreds who share an interest in iron. The International Bioiron Society (IBIS) was founded to advance knowledge about the biological and medical roles of iron and the processes in which it participates with particular reference to those processes of medical and industrial importance. IBIS’s specific objectives are to provide a forum for discussions, to promote research and its publication and advance public education on iron chemistry and biology as well as on disorders due to iron deprivation or excess.

At this year’s meeting, more than 400 scientific and medical professionals attended the four-day event. The delegates shared unique expertise and research findings with likeminded colleagues, all specialists in the field of iron metabolism. Gerry also attended a one-day hemochromatosis patient group meeting that was held at the conclusion of the conference and provides the following report:

“Porto is a beautiful city located on the scenic Atlantic coast of northern Portugal. Known for port wine, its major export, Porto graciously hosted attendees from around the world during this biannual event sponsored by the International Bio-Iron Society (IBIS). More than 400 delegates shared their laboratory and clinical research findings with colleagues. The US delegation included four members of IDI’s Medical & Scientific Advisory Board and 70 other scientists and medical professionals from around the country.

It’s difficult to comprehend the breadth of research that is underway covering all aspects of the science of iron metabolism. Much of the work done in the laboratory, focused on the complex details of mammalian iron regulation using lab animals, and sometimes even using genetically altered animals to mimic human circumstances. Some scientists delve further, into a smaller world that includes the biology of drosophila cells and even bacteria. All of this is important be understand the interaction of iron with the great number on human condition that are affected when normal iron balance is disrupted.

Nearly 300 presentations ranging from slide shows and lectures to poster demonstrations covered all imaginable aspects of iron science. Although the majority of the new research covered discoveries in “lab bench” science, there were a number of good and enlightening presentations describing research involving human subjects. In two hemochromatosis studies undertaken in Denmark, Milman and Pederson reported that only two of twenty C282Y homozygotes found in a cohort of 6,000 Danish men had been treated with phlebotomy, and further that 88% of the untreated men that were tested for iron status demonstrated elevated transferrin saturation and ferritin indicative or clinical iron overload. This research also uncovered 66 male compound heterozygotes (C282Y/H63D), of which 5.4% had both elevated transferrin saturation and ferritin.

The Danish researchers also examined the extrinsic factors that influenced iron uptake in this cohort of men age 30-53. Their findings showed that iron measures were higher in older compared to younger men, and confirmed that high alcohol consumption was associated with higher iron levels. Not unexpectedly, blood donation was significantly associated with hemochromatosis patient organizations. During the meeting, Dr. Gordon MacPhail, M.D., University of the Witwatersrand, South Africa, Arch Mainous III, Ph.D., Medical University of South Carolina; Robert Means, M.D., University of Kentucky; David Meyers, M.D., Kansas University College of Medicine; Mark Princell, M.D., Spartanburg Health-care System. Barry Skinker, M.D., Kansas University Medical Center; Eugene Weinberg, Ph.D., Indiana University; Lewis Wesselius, M.D., Mayo Clinic, Scottsdale, AZ; Mark Wurstler, M.D., Ohio State University

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Acknowledgments:
We especially thank contributors/reviewers, design artists and fact checkers: E.D. Weinberg, Ph.D. (Chair, Publications); Cheryl Garrison, (Editor); Herbert Bontkovsky, M.D.; Mark Wurstler, M.D.; Stephanie Clary; Timm Artus; Jim Hines; Peggy Clark; David Garrison; Gerry Koening; Lee Woods; Chris Kieller; Isabelle Beldel; Jeff Wert; Stephen and Shawn Carpenter.

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Second Quarter 2009
www.irondisorders.org
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Visit our websites www.irondisorders.org www.hemochromatosis.org
IDI Blood Center Focus: United Blood Services

United Blood Services is part of United Blood Systems (UBS), an Arizona based company. UBS serves most of the western United States and is the largest network of blood centers in the US, second only to the American Red Cross. Most recently UBS expanded its variance to include apheresis as a therapy option for hemochromatosis patients. Red cell apheresis, often referred to as double red cell apheresis (DRCA) is especially attractive to patients who have to travel long distances for treatment, or who have issues with needles or dehydration. With apheresis, red cells are removed and plasma and fluids returned to the patient. The needle used in apheresis is a smaller gauge, which some patients appreciate.

Susan Begnaud, Executive Director, United Blood Services of Louisiana comments, “We are very pleased to be able to provide this valuable service for hemochromatosis patients. United Blood Services is dedicated to serving the needs of our communities and this is just one more way for us to be your healthcare partner.”

According to Begnaud, all UBS centers operate under this variance and are able to offer red cell apheresis to HHC patients. Not all centers have the same automated equipment but all of their technology is acceptable for drawing units.

UBS has an excellent website http://www.unitedbloodservices.org/ where you can find the location of centers.

If you live in the Lafayette, Louisiana area and are interested in more information about the hemochromatosis program and services offered, Contact: United Blood Services of Louisiana 1503 Bertrand Drive Lafayette, LA 70506 337-235-5433 x. 3003 Direct Line - 337- 593-7403

Is there a blood center in your area that does not take hemochromatosis blood? Encourage them to call IDI to learn how they can increase their blood supply with an endless resource of donors.

Remembered

Cherished forever, those who paid the ultimate price of Hemochromatosis. Our sincere condolences to their loved ones.

Raymond J. McAuliffe June 13, 2009
James Michael Woltz May 20, 2009

Our Mission

“Iron Disorders Institute exists so that people with iron disorders receive early, accurate diagnosis, appropriate treatment and are equipped to live in good health.”

If you would like for us to honor the memory of someone who is suffering or who has lost the battle with a deadly iron disorder, or you would like to make a prayer list request, call us toll free 888-565-4766 or email Peggy Clark: pclark@irondisorders.org.
Transfusional Iron Overload: Disparities

Thalassemia and sickle cell disease (SCD) patients often require regular red blood cell transfusions. Chronic transfusions can lead to iron overload, a toxic condition of excess iron in the body. Without adequate management, the excess levels of iron will cause progressive damage to the liver, endocrine diseases (diabetes hypogonadism, hypothyroidism, growth failure), and heart, significantly affecting quality of life and survival.

According to Cooley’s Anemia Foundation heart failure is the leading cause of death for thalassemia patients. In the case of sickle cell disease patients, although organ failure or strokes are among common causes of early death, infection, especially upper respiratory is the more prominent cause of death. Although both diseases have anemia and iron overload in common, there is a major disparity in how the consequences and disease potential and in these two groups is approached.

These striking findings were reported by a team of investigators led by Dr. Elliott Vichinsky, Medical Director; Children’s Hospital Oakland Research Institute. Participants included 142 thalassemia and 199 transfused sickle cell disease patients from 35 hematology centers in the US, Canada and London. Investigators reported that endocrine disease is more prominently featured in thalassemia patients compared to SCD patients. Of the 142 thalassemia and 199 SCD transfused patients, outcomes were dramatically different in these two groups:

<table>
<thead>
<tr>
<th>Thalassemia</th>
<th>Sickle Cell Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diabetes</td>
<td>13%</td>
</tr>
<tr>
<td>Hypogonadism</td>
<td>40%</td>
</tr>
<tr>
<td>Hypothyroidism</td>
<td>10%</td>
</tr>
<tr>
<td>Growth failure</td>
<td>33%</td>
</tr>
<tr>
<td>Bone fracture</td>
<td>51%</td>
</tr>
</tbody>
</table>

Fifty-six per cent of Thalassemia had more than one endocrinopathy compared with only 13% of transfused SCD patients. Fung also reported that “transfused SCD patient outcomes did not differ with non-transfused SCD patients, suggesting that the underlying disease may modulate iron-related endocrine injury. In this same group, Fung reports a positive relationship between the severity of iron overload, assessed by serum ferritin, and the frequency of hospitalizations (r = 0.20; P = 0.005). Twenty-three deaths were reported (6 Thal, 17 Tx-SCD) in 23.5 +/- 10 months of follow-up. Within the transfused-SCD group, those who died began transfusion (25.3 vs. 12.4 years, P < 0.001) and chelation therapy later (26.8 vs. 14.2 years, P = 0.01) compared with those who survived. The unadjusted death rate in Thalassemia was lower (2.2/100 person years) compared with that in transfused-SCD (7.0/100 person years; RR = 0.38; 95% CI 0.12-0.99). However, no differences were observed when age at death was considered. Despite improvements in therapy, death rate in this contemporary sample of transfused adult subjects with Thalassemia or sickle cell disease is 3 times greater than the general US population.”

These investigators also noted that Thalassemia patients were “more likely to be screened for iron-related organ injury including an echocardiograph for cardiomyopathy, alanine aminotransferase for liver function and thyroid-stimulating hormone for hypothyroidism. In contrast adult SCD patients, were maintained on simple systematic monitoring of iron and related organ injury in patients with sickle cell disease. She stresses that until the relationship between iron and related comorbidities is better understood, routine monitoring of iron overload in SCD patients who receive transfusions should be considered a standard part of clinical care.”

Dr. Ellen Fung who served as a member of the team on the study comments, “It was true that we observed a much higher endocrinopathy rate in transfused thalassemia patients vs. SCD, but it was confounded by the fact that few SCD patients received transfusions, and were subsequently iron overloaded from an early age which could effect development of endocrinopathy. We attempted to control for these differences statistically, but cannot rule it out completely.”

Dr. Fung continues, “The death statistics in SCD were confounded by the fact that many of our adult SCD patients with severe disease are placed on transfusion therapy to attempt to ameliorate some of the morbidity experienced. Therefore, these very sick patients who died during the study were transfused at a later age, but the relationship between iron overload and death in these subjects was not present as it was with the thalassemia subjects. Our findings were clear that many sickle cell subjects were not receiving the comprehensive care that the thalassemia subjects had been exposed to for years. We hope that our article shed some light on this and will see changes soon.”

RESOURCES:

Glossary of Terms:
www.nlm.nih.gov/medlineplus/
www.cdc.gov/ncbddd/
www.thalassemia.com
www.thalassemia.org
The Role of Iron in Neurotoxicity

Toxicity: the degree to which something is toxic or poisonous
Neurotoxicity can occur because of iron generated free radical formation, lipid peroxidation—and ultimately cell death.

Our brains require huge amounts of iron to function. Because iron is so vital to brain performance it is one the few things that is allowed naturally to cross the blood-brain barrier (BBB), which is a separation of circulating blood and cerebrospinal fluid. Once iron crosses over the BBB, it is transported to sites or areas of the brain by the same transport proteins used in other parts of the body. Transferrin binds with iron and carries it to containment in ferritin. Of all the body systems, our brains and livers contain the largest supply of ferritin.

When the brain needs more iron, transferrin receptors are created. In response, the body increases production of transferrin, which binds to iron and moves it to areas of need. Whenever receptor or transferrin production is faulty, excessive amounts of iron can be dumped into areas of the brain and may overwhelm and impair performance.

Uncontained iron, which is iron not bound to transferrin or stored in ferritin can generate free radical (also called reactive oxygen species or ROS) activity. Although free radical activity is a natural part of the metabolic process, too much of this type activity is harmful resulting in cell death and disease. When this type of activity takes place in the brain and spinal cord, it is called neurotoxicity.

In the neurotoxicity process, portions of the brain are poisoned with too much iron and like all the other systems of the body, the brain function is impaired. Diseases consequential to this type of destruction include (but are not limited to) early onset Parkinson’s, Alzheimer’s, Huntington’s, Lou Gehrig’s, epilepsy and multiple sclerosis. Depression, Friedreich’s Ataxia, pantothenate kinase-associated neurodegeneration (PKAN) and prion disease are also potential consequences of iron mediated ROS.

Also as we age, the iron content of our brains increases. Increased brain iron is present in all the above mentioned neurodegenerative diseases. Alzheimer’s is one of the most prominent concerns for today’s aging population.

The brain tries to protect brain cells by limiting the potential for free iron to trigger oxidation. In response to a central nervous system (CNS) hemorrhage, the brain will overproduce ferritin in an effort to protect astrocytes, cells of the brain critical to normal cell performance.

Another binding protein key to brain health is ceruloplasmin which is a copper based protein necessary to the proper transport and metabolism of iron especially for the brain. If ceruloplasmin is low, iron is not properly managed. Parkinson’s is characterized by an increase in brain iron, but lower levels of ceruloplasmin also correlate with early onset Parkinson’s.

Iron accumulation may be a predictor of the duel presence of Parkinson and Alzheimer’s. The development of PD during the course of AD appears to be associated with the accumulation of iron in the brain.

Wistar rats are often studied before studies on humans. How the rat behaves in a maze is observed and reported. In one study, rats fed moderate doses of iron for 5 consecutive days performed poorly in the maze; they exhibited diminished spatial learning and emotional behavior with hampered proficiency in the maze. The excess supplemental iron overwhelmed the normal mechanisms that shield the brain from iron toxicity.

Harmful amyloid plaques which can collect in delicate blood vessels in the brain are strongly enhanced by the presence of iron. Iron chelation has been suggested as a potential therapy for this and other types of neurodegenerative disease. However the current chelators cannot cross the blood brain barrier. When more efficient iron chelators are discovered, they will need to be fine-tuned to remove excess/misplaced iron while leaving metabolically useful iron intact.

Resources:


Image: Don Carstens Medical Imaging Artville
Western Regional Hemochromatosis Conference: Reno, Nevada

The Presenters

John A. McDonald, MD, PhD; Vice President, Division of Health Sciences, University of Nevada, Reno welcomed attendees.

Lewis J. Wesselsius, MD, Chief, Pulmonary and Critical Care Medicine, Mayo Clinic, Scottsdale, Arizona and IDI Advisory Board Member chaired the event and spoke about dangers of inhaled iron.

Gordon D. McLaren, MD; Professor, Department of Medicine, University of California, Irvine; VA Long Beach Healthcare System Long Beach, CA provided an overview of HHC and HEIRS.

Joanne M. Jordan, MD, MPH, Associate Professor, Medicine and Orthopaedics, University of North Carolina School of Medicine, Chapel Hill, North Carolina spoke about HHC arthropathy.

Suchi Pandey, Assistant Medical Director, Blood Centers of the Pacific spoke about the FDA variance for blood centers.

Ralph G. DePalma, MD, FACS, Director of Transplant Services, Department of Veterans Affairs, Professor of Surgery, Uniformed Services UHS, Washington, DC spoke about the history of phlebotomy.

During the break, patients mingle and network. Hemochromatosis patient Harry Kieffer talks with attendees about the challenges of hemochromatosis arthropathy.

Attendees listen intently to presenters.

Attendee enjoys handouts.

Medical professionals share impressions of the event and educational materials.

Patients Rick Kaufmann, Veterans’ Hospital, Reno; Lynn Scally, Public Works Dept., Santa Cruz, CA, Gerry Koenig and Pam Reese, Reese Realty, Reno, NV share their unique experiences with hemochromatosis-iron overload.
Comments were warm and supportive from clinicians and patients. One Lake Tahoe, CA gastroenterologist, remarked “I learned more in these few hours than I did in a two-day (gastro) seminar.”

“I wanted to thank you again for bringing the HHC seminar to Reno. It was so incredible for me to talk to you, the medical experts and others with hemochromatosis. Since I was only diagnosed about five months ago, I have much to learn and understand. I’ve been trying to figure out my health problems for about 20 years and finally feel like I’m getting somewhere. Your program helped me immensely!”

–Hemochromatosis patient, Lake Tahoe, CA

“I am blown away by the (IDI) materials and level of professionalism.”

---blood bank special collections supervisor.

“Good, professional materials.”

–Hemochromatosis patient Yorba Linda CA

“I feel more able to help myself now.”

—Hemochromatosis patient, Willits, CA

“It was refreshing to have doctors listening to patients, asking questions and being honest about answers when they may not really have one!”

–Hemochromatosis patient, Willits, CA
DRCA: The “Cadillac” of phlebotomy procedures
The Stephen Carpenter Story
Part I

Perhaps the cruelest thing about being diagnosed with hereditary hemochromatosis at age 41 was the fact that once the diagnosis finally arrived, my first two hematologists seemed to think that their work was done. In reality my struggle for treatment had only just begun – how to get the iron out?

I was grateful for the diagnosis after 10 years of failing health - but when it came to actually removing the poisonous excess iron from my body through blood donation, I found good information hard to come by. For nine months my symptoms worsened as I flailed around, searching for an alternative to the medieval process of bloodletting. Not only did I have a basic fear of needles, I had a history of difficult IV insertions and nurses and didn’t “like” my left arm at all. I was also concerned that repeated punctures in my “good” right arm over the next year or so might leave my veins damaged for the future draws that I would need for the rest of my life.

My first two doctors’ only advice on dealing with my strong fear of needles was to “get tough and go give blood.” Though they listened skeptically to my quest for other vein-friendly options for removing the excess iron, and said they’d get back to me, they eventually abandoned me to my illness and my fears, untreated. I even tried to buy leeches as an alternative, but my illness and my fears, untreated. I even tried to buy leeches as an alternative, but found that the good ones were endangered, expensive, and good for only one use! As my cardiac symptoms worsened, I was well on the way to suicide-by-iron-overload.

Had it not been for the Internet, which is how I found the Iron Disorders Institute, I might never have learned of the simple but brilliant iron-removal procedure which my doctors had previously discarded, called Double Red-Cell Apheresis, or DRCA. If you have iron overload, are having a hard time getting used to the idea of having someone repeatedly stick a metal tube into your arm to regain your health (a very unnatural thing!), and want an easy alternative to the standard prescription of giving blood until your arm feels like a pin cushion, then - take it from a needlephobe – DRCA is your ticket.

With one small needle, and only half the number of “sticks,” DRCA is the “Cadillac” of phlebotomy procedures. A special machine siphons off a double-shot of iron-rich red blood cells while returning the rest of the good stuff to your body, resulting in less fatigue. DRCA is offered at many local blood banks but not widely publicized; as a needlephobe, I wholeheartedly endorse it. It has put me on the road to recovery and, in many parts of America, is free of charge!

The people at the Iron Disorders Institute connected me with local resources; a hematologist James L. Cole and United Blood Systems, a treatment center that offered DRCA.

Treatment couldn’t have come too soon. By the time that I was finally diagnosed in August 2008 with homozygous hereditary hemochromatosis, mutation C282Y/C282Y, the iron overload was making my heart misfire, and my shortness of breath was so severe that I was often too weak to walk to the mailbox. In the year before I demanded an iron test, after worsening chest pains and a fluttering heartbeat, I quietly told a couple relatives, “I don’t know what’s wrong with me, but I feel like I’m going to die.” I had suffered from a growing variety of seemingly random symptoms since my late 20s, including tiredness, lethargy, profuse sweating in cool rooms, stiff and painful joints, abdominal pain, elevated liver counts, mental fog, irritation, psoriasis, tinnitus and vertigo. Several doctors told me to see a psychiatrist, and family members had been whispering, “what’s wrong with Stephen?” Two doctor friends joked that I was going through “male menopause.” One doctor found my search for a solution so seemingly obsessive that she wrote in my chart that I was “overly somatically aware.”

At this point my ferritin level was nearly 1,600, my transferrin saturation percentage was 88%, and now I was officially ill. My iron-rich Southern diet of oysters, shrimp, beef, liver, one-a-day vitamins and cooking in iron skillets certainly hadn’t helped. As bad as I felt, I could see little hope of any future, with my health ruined. But, for my family’s sake I decided to try. I am here to tell you that DRCA does work, and that partway through the process, removing the iron works wonders. Would you believe me if I told you that after four DRCA treatments I felt like I was 25 again?

So what is DRCA? Double Red-Cell-Apheresis is really nothing more than two blood donations at once – but with some big advantages. The unit is much like a dialysis machine- it painlessly draws blood, spins it up to remove the red cells, which then can be given to cancer and leukemia patients, and then returns the rest of the plasma through the same small single needle. It takes about 20 minutes and because the plasma is returned, one doesn’t get that immediate weakness and dehydoration as with regular whole-blood donation. Plus, twice the amount of iron-rich red cells are taken at once, which means that over the course of the de-ironing process, only half the number of needle sticks are needed! Finally cool, too, to watch the red stuff go out and the clear stuff go back in as the wheels spin. Whoever invented DRCA was a genius!

How did I psyche myself up to try it? First, the needle is smaller than regular donation. Second, my doctor prescribed two items to make everything easier – a tube of Lidocaine 4% cream to rub on my arm and a small 1-mg. Xanax tablet, to be taken 30 minutes before the treatment. I can never even tell when the needle goes in. This dosage really works because it keeps me and the phlebotomy nurses relaxed and then a family member can drive me home.

Who is eligible for DRCA? Anyone in basically good health can donate using the DRCA machine every 112 days, free of charge. Hemochromatosis patients may also donate, free of charge, on a doctor’s order of “therapeutic phlebotomy,” and as often as every two weeks or more, as long as a painless “finger-stick” shows that their hematocrit number is safely high enough, meaning that their blood is rich enough in red blood cells.

My goal is to continue the regimen of one DRCA every two weeks, with my minimum allowable hematocrit set by my doctor, until the hematocrit is slow to recover, then to let my body recover for 4 weeks. Rinse and repeat until the hematocrit stops recovering, meaning that the stored iron overload is gone, and then go to maintenance therapy four times a year. While I still have a year or so to go, and though the DRCA treatments take me through energy highs and lows as the iron-rich red blood cells are periodically removed, allowed to re-grow, and then removed again, my cardiac symptoms have disappeared and my breathing improved with the removal of iron.

I have already had a taste of what life has in store - during a recent 4-week “sabbatical” to allow my red cells to rebuild, my renewed energy was incredible, I was clear-headed and energetic as I can’t remember since my 20s. I was able to work on my old trucks again, sang and played guitar in a club, and even went dancing recently with some nursing students down from Shreveport! This was just a brief taste of what life will be like without iron overload – like 100 pounds had been lifted off my back, and I am looking forward to next year when this is done. Thanks to DRCA, “look out, World, here I come!”

Part II of Stephen Carpenter’s story will appear in Fourth Quarter 2009 nanograms. In part II Stephen will share details of his experience with an understanding physician, importance of hematocrit, MCV, problems and successes related to his treatment.
Q: Is there a genetic relationship between hemochromatosis and celiac disease? What is the percentage of overlap between the two?
A: Yes, there appears to be an overlap of Celiac and Hemochromatosis however the exact percentage is not known. Due to the increased absorption of dietary iron a person with Hemochromatosis may actually help the Celiac to be less anemic. Both Hemochromatosis and Celiac disease are of genetic origin, meaning that it runs in families. Celiac disease is both a disease of malabsorption—meaning nutrients are not absorbed properly—and an abnormal immune reaction to gluten.

Celiac disease is also known as celiac sprue, nontropical sprue, and gluten-sensitive enteropathy. Sometimes the disease is triggered—or becomes active for the first time—after surgery, pregnancy, childbirth, viral infection, or severe emotional stress. Celiac disease affects people in all parts of the world. Originally thought to be a rare childhood syndrome, celiac disease is now known to be a common genetic disorder. More than 2 million people in the United States have the disease, or about 1 in 133 people. Among people who have a first-degree relative—a parent, sibling, or child—diagnosed with celiac disease, as many as 1 in 22 people may have the disease.

Celiac disease is also more common among people with other genetic disorders including Down syndrome and Turner syndrome, a condition that affects girls’ development. As far as Hemochromatosis 1 in 7 persons has one gene and 1 in 200 has two genes. A person with 1 or 2 genes may not have any symptoms, yet, may be at risk for the disease and that is why it is so important to do the 3 blood tests at least once a year: Serum Ferritin, Serum Iron, and the Serum Transferrin Saturation Percentage. A complete blood count may also be useful. Do not eat for 12 hours before the blood tests, you may however drink some water.

For more information on Celiac Disease: American Celiac Disease Alliance 2504 Duxbury Place Alexandria, VA 22308 Phone: 703–622–3331 Q: Email: info@americanceliac.org Internet: www.americanceliac.org

What damage will occur to our veins over years of Phlebotomies?

It is really an individual case by case... Some people long-term may have more damage to their veins than others. Veins can roll or collapse with repeat phlebotomy, especially if numerous blood donations are needed to bring iron to normal levels. There are some alternative therapeutic approaches for patients with extremely high iron. These include: chest port, double red cell apheresis or combination iron chelation. These are exceptional approaches and not recommended for those who can tolerate phlebotomy and should be discussed with your physician. If you have a skilled phlebotomist you may experience very little damage. The way to assist yourself is to make inquiries at the lab you visit for the best phlebotomist, and then ask for that one each time.

Before a phlebotomy, it may be helpful to move the needle site often, keep warm, drink fluids, walk, and move your arms.

What is your Question?
One way for new patients to learn about iron disorders is from other patients on the Excess Iron Discussion List. Join NOW!
If you need help getting on the LIST send us an email. SClarkey@irondisorders.org

If we’ve helped you, please help us by donating.
BECOME A Volunteer and a MEMBER With your membership dues you will receive your handsome IDI membership pin and a printed copy of nanograms.
For details about membership please call Peggy Clark, Member Services Coordinator 888-565-4766; email: pclark@irondisorders.org or visit our websites: www.irondisorders.org and www.hemochromatosis.org

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Misunderstood—Misinformed

Pitfalls of “Do It Yourself” Diagnosis and Treatment

Patients are encouraged to take an active role in getting a complete diagnosis of hemochromatosis and appropriate therapy for excess iron reduction. In today’s Internet Direct to Consumer product availability, patients can have DNA tests, lab work entirely without involving their healthcare provider. Odd symptoms and abnormal liver tests can cause a doctor to be suspicious and wrongly label a hemochromatosis patient as a drinker or mentally unbalanced. This fosters a level of mistrust and attitude of defiance in the patient.

Once a patient feels mistrust or thinks that the doctor lacks knowledge, the probability for self diagnosis and treatment is highly likely. As determined patients press on without the help of a physician, some do in fact reach a complete diagnosis and learn the best way to address excess iron levels. Others do not have such a positive outcome. They can become far too anemic, go to extremes with the diet and supplements and find themselves facing irreversible consequences.

Being proactive is imperative for the hemochromatosis patient. Knowledge gaps do exist in the medical community not just about hemochromatosis but about other iron disorders such as iron deficiency and anemia of inflammatory response. Patients can be a helpful resource to a doctor who has had little experience with hemochromatosis.

Doctors appreciate the resources provided by Iron Disorders Institute (IDI). The IDI Physician Hemochromatosis Reference Chart gets high praise from medical professionals. At the Western Regional Hemochromatosis Conference one half-day was devoted to clinicians, all of who expressed a positive evaluation of IDI’s educational materials. The diagnosis algorithm was among the favorites of attendees; the sample order, treatment, genetics and diet pages all received positive remarks.

These tools are available to patients for their doctors FREE of charge online and through IDI’s Patient Services. In parting I encourage you to get a copy of this reference chart and share it with your doctor; it has been my experience that in doing so, both the doctor and the patient will benefit!
The EGG is one of nature’s most ingenious foods when it comes to iron balance. Although eggs contain a good bit of iron, they also contain a proteins that protect us from harmful invaders and one that inhibits iron absorption! Lactoferrin contained in the whites of the egg protects us from harmful invaders and phosvitin a phosphoprotein lowers the bioavailability of iron. The iron inhibiting ability of eggs is called the “egg factor”. The egg factor has been observed in several separate studies. One boiled egg can reduce absorption of iron in a meal by as much as 28%! In a recent Danish study of men who regularly consumed eggs had lower serum ferritin levels.

**Steps for planning a meal to balance your iron intake**

- Estimate the amount of heme (animal source) and non-heme (plant source) iron in your meal.
- Determine what to substances to add or substitute to improve iron absorption—**if you need more iron**, or impair iron absorption—**if you need less iron**. See the list below of substances that improve or impair iron absorption.
- Plan ahead! If you plan your menus in advance and use a shopping list, you will be less prone to impulsive eating and processed foods.

*Get a FREE copy of the Iron Disorders Institute (IDI) MENU PLANNER FORM. You can also download this form from our website [www.irondisorders.org](http://www.irondisorders.org)*

*If you are a member,* you can request we send you a printed copy. Call us toll free: 888-565-4766!

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### Recipes for Iron Balance

#### Quiche

**Crustless Quiche**

- 3 eggs
- 3 slices turkey bacon
- 3 slices Canadian bacon
- 1 small onion (finely diced)
- 1 cup grated Swiss cheese
- ½ cup milk
- Handful of fresh spinach leaves chopped finely
- Salt & Pepper to taste

Preheat oven to 400. Spray the bottom and sides of a pie plate with olive oil spray. Place onion in a microwave safe bowl, sprinkle with water and cover with a paper towel. Microwave on high approximately one minute or until tender. Cook both turkey and Canadian bacon and drain well. Crumble the turkey bacon and chop Canadian bacon.

In the bottom of pie pan, layer the onion, chopped spinach, and bacon. Cover with grated cheese. In a separate bowl, mix eggs, milk, salt and pepper together until frothy. Pour egg mixture over ingredients in pie plate. Be careful not to fill to high or it will boil over. Cook on 400 for 15 minutes, then turn down to 350 and cook approximately 30 minutes or until toothpick comes out of center clean. If top gets to brown you can cover with a foil tent.

*This recipe is low in fat, low in heme and non-heme iron and high in protein—a great way to get started in the morning!*

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**FOODS AND SUBSTANCES THAT IMPROVE Iron Absorption**

- Beta-carotene
- Sugar
- Acidic foods or beverages
- Alcohol
- Vitamin C supplements
- Red meat

**FOODS AND SUBSTANCES THAT IMPAIR Iron Absorption**

- Coffee
- Tea
- Eggs
- Fiber
- Chocolate
- Calcium supplements

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Second Quarter 2009
ABOUT IRON
Iron is mineral that we get from food. All living things must have iron to survive. Humans need about 1 milligram of iron a day to have enough energy to function. People lose about 1 milligram of iron per day in sweat, skin flakes or tears. Most people get enough iron from the diet, but some have Iron-Out-of-Balance™. This is any condition where iron levels in the body are not normal.

TESTS TO DETECT
Iron-Out-of-Balance™ is detected with blood tests. The most common tests include:
- serum iron
- total iron-binding capacity (TIBC)
- serum ferritin
Other tests or procedures are needed to determine the cause of Iron-Out-of-Balance™ examples include complete blood count, retic count, B12 or folate, genetic testing, liver biopsy, and bone marrow aspiration. Our books are excellent resources for understanding iron disorders such as hemochromatosis, anemia of chronic disease, iron overload with anemia and iron deficiency.

IN YOUR GENES?
Many iron disorders are inherited; that means it’s in your genes. If you are diagnosed with an inherited iron disorder, even if you are just a carrier, be sure to tell all your blood relatives: your parents, brothers and sisters, cousins, aunts and uncles. They need to know; if it is in their genes too, knowing might save their life!

THREE VIEWS OF IRON
IN USE: determined by measuring hemoglobin.
BEING TRANSPORTED: determined by measuring serum iron and TIBC (total iron-binding capacity)
CONTAINED IN STORAGE: determined by measuring serum ferritin.
YOU NEED ALL THREE VIEWS for a complete picture of your iron levels.

<table>
<thead>
<tr>
<th>Age Group</th>
<th>Hemoglobin Ranges (Normal Range)</th>
<th>Hemoglobin Ranges (Adolescents, Juveniles, Infants &amp; Newborns)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adult Males</td>
<td>13.5-17.5 g/dL</td>
<td>12.0-16.0 g/dL</td>
</tr>
<tr>
<td>Adult Females</td>
<td></td>
<td></td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Age 6-18 years</td>
<td>10.0-15.5 g/dL</td>
<td>10.0-17.0 g/dL</td>
</tr>
<tr>
<td>Age 1-6 years</td>
<td>9.5-14.0 g/dL</td>
<td>12.0-20.0 g/dL</td>
</tr>
<tr>
<td>Age 6 mos-1year</td>
<td>9.5-14.0 g/dL</td>
<td>Newborn 14.0-24.0 g/dL</td>
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<td>up to 300ng/mL</td>
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<tr>
<td>Male ages 10-19</td>
<td>23-70ng/mL</td>
<td>Infants 7-12 months 60-80ng/mL</td>
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<tr>
<td>Female ages 10-19</td>
<td>6-40ng/mL</td>
<td>Newborn 1-6 months 6-410ng/mL</td>
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<td>Children ages 6-9</td>
<td>10-55ng/mL</td>
<td>Newborn 1-30 days 6-400ng/mL</td>
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<td>Children ages 1-5</td>
<td>6-24ng/mL</td>
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Important Ranges
Hemoglobin measures the amount of iron in the blood that is carrying oxygen to vital organs. Hemoglobin will be within normal range unless you are iron deficient or have anemia of chronic disease.
Ferritin (serum) is a measure of contained iron. Ferritin will be elevated if you have too much iron in your body or if you have inflammation. Ferritin will be low if you are iron deficient.

Important Ferritin Reference Ranges

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Important Ferritin Reference Ranges

- Serum ferritin
- Total iron-binding capacity (TIBC)
- Serum iron

THERAPY TO CORRECT
People with normal hemoglobin and high body iron can have therapeutic phlebotomies. If they cannot tolerate the phlebotomies, they may be candidates for iron chelation therapy. This form of therapy is usually used with iron overload patients who are anemic. Iron chelators are pharmaceuticals that will specifically bind to iron.

DYK?
If a pregnant woman’s iron stores (ferritin) are adequate, taking unnecessary iron supplements in amount >100mg could affect hemoglobin concentration, premature and low birth weight babies. Read more about iron during pregnancy in the next issue of nanograms.

C.R. Hume says, “Let us help you remain... IRON SMART!”
C.R. Hume is IDI’s mascot; he is a health-minded ferret, who helps raise awareness about the benefits of blood donation. His name is suggestive of “serum”. Together, his name and genre sound like serum ferritin.
Our mission: Iron Disorders Institute exists so that people with an iron disorder receive early, accurate diagnosis, appropriate treatment and are equipped to live in good health.

Iron Disorders Institute publishes books about iron for patients and physicians.

BECOME A MEMBER OF IDI

and we will send you a book of your choice!

JOIN online at www.irondisorders.org

www.irondisorders.org

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