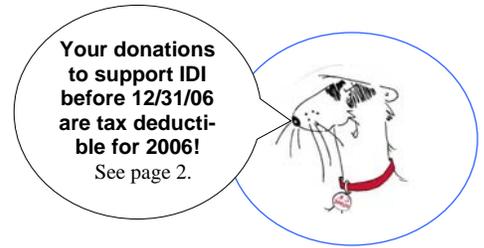


“The value of a man resides in what he gives and not in what he is capable of receiving.”

Albert Einstein, physicist



IDI's way of keeping in touch with you!



NOVEMBER/DECEMBER, 2006

IRON DISORDERS INSTITUTE

VOLUME 6, ISSUE 6



As Executive Director, I am extremely privileged by this opportunity to introduce the dedicated individuals who are the bulwark of Iron Disorders Institute National Headquarters in Greenville, South Carolina. Our staff and volunteers unstintingly respond to a myriad of telephone, email and written requests from patient and their families by locating iron-related health information, developing and updating web site information, distributing IDI literature and publications, not too mention supporting IDI's national volunteer organization.

Beginning with this issue, we will profile a staff member and a volunteer in each issue of the newsletter. See our staff and volunteer profiles on page 3.



IRON AWARENESS FOR THE MENOPAUSAL OR NON-MENSTRUATING WOMAN

Reprinted from *idInsight*, Spring 2005

Loss of period = loss of benefit for prevention of disease

As a woman grows older, her iron needs change. Loss of menstruation, changes in hormone levels and the perhaps the increase in chronic disease all factor into the mature female's lesser need for iron.

With the loss of menstruation, which usually occurs for females in their mid-fifties, she will no longer be losing iron every month. This regular iron loss through blood loss is now likely to be retained in the tissues of liver, heart, joints, pancreas and anterior pituitary. The excesses in these vital tissues if not addressed can lead to osteoporosis, cirrhosis, heart attack and diabetes. Signs and symptoms generally begin to appear 10-15 years post-menopause or after the period stops.

Among the women of childbearing age, more and more are being urged to take oral contraceptives to reduce the frequency of monthly bleeding or to regulate periods. Rather than iron loading 10-15 post menopause, these women will begin to get overloaded in their 20's and 30's just as men do. Stopping of one's menstrual cycle for whatever reason can result in iron overload.

For the first decades of a woman's life, one message is constant "Get plenty of iron!" Females who experience heavy

menstruation or do not eat much red meat are at increased risk to become iron deficient, but not all may require supplemental iron. Taking supplemental iron without laboratory evidence of iron deficiency can be dangerous even though this practice is rather commonplace in the USA.

Many of us are conditioned to take iron if we are tired. The Baby-Boomer generation is very familiar with the Geritol product, a potent iron supplement marketed through commercials with the message: "Tired Blood? Take Geritol! These jingles became so ingrained in our culture that some physicians may continue to recommend supplemental iron to patients complaining of fatigue without a complete iron panel to determine body iron levels.

Body iron levels are best determined with a complete blood count, fasting serum iron, total iron binding capacity and serum ferritin. Among these tests, serum ferritin is important for distinguishing between iron deficiency anemia, anemia of chronic disease and iron overload.

Since chronic fatigue is often reported by older women, these

See **Menopause** continued on the last page.

<p>THE GENETIC COUNSELING CORNER LISA KESSLER PAGE 2</p>	<p>I AM A FAMOUS PERSON WHO DIED OF HHC PATRICK MCKEEVER PAGE 2</p>	<p>YOUR DONATIONS SUPPORT IDI 2006 PAGE 2</p>	<p>STAFF & VOLUNTEER PROFILES PEGGY CLARK & CAROL JORDAN PAGE 3</p>
<p>ANNA MARIE'S LEGACY THE JIM HINES STORY AUTOPSY, GENES & HHC PAGE 4</p>	<p>A LETTER FROM THE AFTERLIFE TO MY LOVED ONES PAGE 7</p>	<p>THE RUSTY CURMUDEON JIM HINES PAGE 10</p>	<p>HOLIDAY GIFT SUGGESTIONS THE IRON STORE PAGE 10</p>

IDI's mission is to reduce pain, suffering and unnecessary death by disorders of iron through education, awareness and facilitating research

THE GENETIC COUNSELING CORNER

By Lisa Kessler

**REACHING OUT**

Genetic counseling is defined as “the process of helping people understand and adapt to the medical, psychological and familial implications of genetic contributions to disease. This process integrates the following:

- Interpretation of family and medical histories to assess the chance of disease occurrence or recurrence.
- Education about inheritance, testing, management, prevention, resources and research.
- Counseling to promote informed choices and adaptation to the risk or condition” (National Society of Genetic Counselors, 2006).

As many of you may be aware, genetic counselors have training to address both the genetic and psychosocial aspects of different conditions. Our last column focused on some genetics terminology and the discovery of the *HFE* gene. In addition to the medical genetics focus, I am most passionate about learning about the impact that a diagnosis has on an individual and his/her family.

As you know far better than me, a diagnosis of hemochromatosis can affect the whole family, not only the individual who is diagnosed. These affects can be both positive and negative. In genetic counseling, we often say our patient is the family. What I don't always know best as a genetic counselor is the most effective way to help patients share information about a diagnosis with their family members. I provide guidance, role-play, and often help patients by writing family letters to share the information with other family members. DNA Direct also recognizes the value of sharing information with family members and provides family letters for those who test with us. Some patients have told me that they felt a great deal of pressure to be the one in their family to have to explain all of this new genetic information, while others felt empowered by this role.

Why share information about a diagnosis of Hemochromatosis with your family members? One of the main reasons is because an individual's diagnosis may mean that family members have an increased risk to develop hemochromatosis. These risks vary depending on the relationship. Click on the link for our resource center to learn more specific risk information: Hemochromatosis Risk in Families:

http://www.dnadirect.com/resource/conditions/hfe/GH_Hemo_Risk.jsp

How did you share the information with your family members about your diagnosis? Let me know what worked best and with your permission, share your experiences with this community so that we can all learn from your success. As always, feel free to contact me with any genetics questions at:

lkessler@dnadirect.com

Or

1-877-321-0077

*Lisa Kessler is a certified genetic counselor with DNA Direct;
<http://www.dnadirect.com>*

I AM A FAMOUS PERSON WHO DIED OF HH

By Patrick L. McKeever

I am a famous person who died of HH.
You all know me very well or have at least heard of me.

I am your family member who died from HH.

I am the spouse, mother, father, uncle, aunt, son, daughter, cousin or married relative that died of HH and its complications.

I am very real in your life, you lived with me, I raised you, and you watched me suffer and die or you knew or heard of me through the family.

Yes, I died of HH and HH complications.

At that time not much was known about HH. Today, there is much more awareness, publicity, education, support and discussion of HH. You are all doing a great job.

I read these posts daily and over the past five years the medical information explosion and publicity of HH has been exponential. Keep up the good work.

Worry not about me but do worry about your children and family members. Without education, support and encouragement they too will suffer from HH and its complications.

Keep up the good work and take care of future generations.

Thanks to all who are doing research on HH and iron related disorders.

Your web sites and on line support groups are wonderful.

Thanks to all on this list for your continued efforts and generous contributions. You are making a difference!"

Your Loved One

Editor's Note: Patrick originally posted this poem on IDI's Excess Iron List. Patrick has given permission to use this poem to advance iron awareness.

***An end of year donation makes a difference.
So, please, be generous this holiday season.***

The generosity of our donors has been and always will be important as we continue to provide vital services for patients, their families and physicians.

Consider your donation to Iron Disorders Institute an investment in the health of future generations.

Please make a contribution today!

You can obtain more information from our web site:

<http://www.irondisorders.org/Donate/>

IDI's staff and volunteers thank you for your continued generosity and wish you and your loved ones a happy holiday and very healthy New Year.

Your contributions are fully tax-deductible to the amount allowed by law.

Iron Disorders Institute National Headquarters Staff and Volunteers

Iron Profile: Peggy Clark, Executive Assistant

“Before coming to work at IDI, I had never heard the word hemochromatosis let alone how to spell it”

Peggy Clark is the voice you hear when you call IDI, in fact she is Patient and Support Services. “Before coming to work at IDI, I had never heard the word hemochromatosis let alone how to spell it”, said Peggy.

“My work with IDI has been a whole new adventure for me. I am still in the learning process for all of the different iron related disorders. I hope it is true that: ‘You can teach an old dog new trick.’ Cheryl and David have made me feel like a part of their family. In a work environment there is no better feeling than to be appreciated and respected.

I wish everyone could sit at my desk for one day to answer the phone or talk with the people who walk into this office. When patients first contact us, they are often scared and very worried. Sometimes all it takes is just listening, other times the name of a doctor or one of our fact sheets is enough to change their fear and worry to relief. It is rewarding to bring a smile to that person, but also gratifying when they call us back to say thanks for the help and support”

Peggy said the closeness she feels with the entire IDI staff became apparent when tragedy struck on July 6, 2006. That was the day Peggy’s 19-year-old son, Jason, an IDI volunteer with a “Hollywood smile,” was killed in a car accident. Jason’s passing left Peggy, her husband of 31 years and their 21-year-old daughter facing one of life’s most difficult challenges.

“I’m grateful for the support of my co-workers,” Peggy said. “Things might be different from now on, but in my heart I’ll always have two children.”

Peggy was raised in Cheraw, SC, three hours east of Greenville. She has always enjoyed working with people. Her previous employment was as secretary of the First Presbyterian Church in Cheraw and as a Customer Service Representative for South Carolina National Bank (now known as Wachovia Bank.)

Peggy is the hub of IDI’s operations. Pictured here, she is filling patient requests for IDI brochures, books and materials. Requests mostly arrive at national headquarters by phone and email.

**Iron Profile: Carol Jordan, Web Support**

“The Internet was still quite new and information about “rare disorders” was quite difficult to obtain and more difficult to understand.”

Carol is IDI’s newest “Web Nanny.” She surfs the Internet in search of iron myths, iron information and websites that would benefit from a relationship with IDI.

Carol was told in the late 1980s that her ferritin was a little high. It was thought that there was nothing to be concerned about because she was still in her early 30s. At that time many physicians believed that a woman, particularly one in that age group, would be iron deficient and not iron overloaded. The Internet was still quite new and information about “rare disorders” was quite difficult to obtain and more difficult to understand.

Only after experiencing severe fatigue, joint pain, frequent infections, and an ever-increasing ferritin level was there any concern that these problems might be related to the presence of too much iron. The concern that there might be too much of a good thing going on with Carol was first voiced by her mother, Estelle Cobb. She had read an article about a college football coach with a disorder called hemochromatosis. She sent for information on the disorder and became convinced that this was the cause of Carol’s ever-increasing problems. In October 1992 more extensive testing revealed a tentative diagnosis of hemochromatosis. Fortunately, approximately a dozen phlebotomies were required to de-iron her sufficiently. Most of the fatigue and joint pain was relieved shortly after beginning phlebotomies.

In 2005 Carol was given the opportunity to have DNA testing through an agreement with DNA Direct and IDI. The result of homozygous C282Y mutation was actually quite a surprise. While Carol’s father died at 63 of liver and pancreatic cancer, her mother lived until she was 89 with few, if any, iron related problems. She passed away before Carol received the DNA test results. There are multiple iron-related cancers in Carol’s father’s family including lymphoma, multiple myeloma and Hodgkin’s lymphoma.

Carol lives outside of Atlanta with George, her husband of 28 years, and their cat. They have three daughters and seven grandchildren. While not surfing the web for IDI she works in an accounting practice for the best boss in the world, enjoys crafting, the beach and gaining new knowledge.

Editor’s Note: Carol is one of many volunteers that volunteer their time, effort and experience electronically. In effect, volunteers like Carol are “televolunteering.” This is akin to telecommuting.

ANNA MARIE'S LEGACY: THE JIM HINES STORY

Editor's Note: Jim Hines' personal story first appeared in the Spring, 2004 issue of IDI's magazine, *idinsight*. IDI staff thought that this particular column would share insights to how an autopsy of a loved one can positively impact the health and well-being of future generations.

"When one door closes another door opens; but we often look so long and so regretfully upon the closed door, that we do not see the ones which open for us."

Alexander Graham Bell (1847 - 1922)

No parent should endure the death of child. It's not in the natural order of things. Yet our mother had to bear the unnatural grief which comes from the loss of her oldest daughter at an early age. Our entire family grieved when Anna Marie passed away on September 4, 1975. Anna Marie was 28.

Anna Marie Bennett Hines was born prematurely, as a result was always very fragile. Throughout her short life she endured frequent illnesses and several medical conditions: surgery of the mastoid and epilepsy, to name a few. Early on, Anna Marie experienced several grand mal episodes followed by many years of petite mal incidents. She was under constant medical care, which helped minimize and control the effects of her epilepsy. Nevertheless, Anna Marie remained a very positive and determined individual, accepting the limitations placed on her by her



medical conditions.

Although she had missed several years of school because of illness, her determination was rewarded when she graduated from high school in 1969 at age 22. Anna Marie considered this her personal, major achievement. She subsequently went to work in the local public library where her pleasant personality and hard work endeared her to all the librarians. One thing you could count on: Anna Marie always had a smile and a good word for everyone that came in contact with her. Physically, Anna Marie was tiny: 4' 9" and less than 100 lbs, yet in stature she stood as tall as an oak.



In late August, 1975, Anna Marie was admitted to a local hospital with what was thought to be a viral type illness. Six days later she was transferred to a Boston teaching hospital with a diagnosis of *azotemia*. She died 2 days later with a clinical diagnosis of suspected *myocarditis* and *cardiopulmonary* failure along with other complications; such as *hypoxemia*, *disseminated intravascular coagulation*, *azotemia*, and *anasarca*.

Our mother, although experiencing extreme grief over the loss of her oldest daughter, companion and sometime confidant, had the wisdom and foresight to request an autopsy be performed. Her motivation was initially very simple: someone should learn from this tragic loss of life, which had occurred at an early age. Little did we realize at the time that even in death Anna Marie would be teaching her family. The true meaning of her legacy would not be fully appreciated until many years later.

The provisional autopsy findings (See Exhibit 1) were consistent with the clinical diagnosis with some discussion of alcoholism due to the condition of her liver. Our mother went ballistic at the mere mention of alcohol. Alcohol had never passed Anna Marie's lips! At her insistence, the pathological findings were revisited.

The final autopsy report (See Exhibit 2, next page) was quite a shock to us all because it indicated the major cause of death as *hemochromatosis*. The autopsy summary indicated "extensive iron depositions" in most of her organs. The pathologist noted in his report that absent a family history of excessive iron overload, he could only infer a pathological diagnosis of *idiopathic hemochromatosis* based on "considerable resemblance."

What was this thing called *hemochromatosis*? Our mother was determined to find out what had killed her daughter prematurely.

Fortunately, she lived in an area where public and medical school libraries were abundant. The internet was not available at the time; consequently our mother had to do her research the old-fashioned way: one library at a time, one medical journal at a time, one article at a time. After many hours of driving to and from home, countless telephone calls, and reading dusty and out-of-date medical journals our mother's persistence paid off. Reference to *hemochromatosis* was scant in most of the medical journals. Nevertheless, she had collected sufficient information within a little more than a year to realize the hereditary significance of Anna Marie's diagnosis. As a result of her perseverance and tenacity she concluded that her remaining children were also at risk.

Our mother immediately insisted that my 2 other sisters, Kathy and Irene, have their iron levels tested.

Kathy's iron levels were within normal ranges. My other sister, Irene was pregnant; consequently, her doctor delayed any testing until September 1978, 6 months after my niece was born. The doctor's felt that it would be best to delay iron testing until her body chemistry and hormonal levels returned to normal: changes resulting from pregnancy might skew the results. Ultimately, the results of her blood tests

See **Anna Marie** continued on next page.

Date of Autopsy 9-5-75

Decreased consistency of myocardium consistent with cardiomyopathy

Cardiac failure (clinical)

Biventricular dilatation

Pulmonary edema

Passive congestion, liver, slight

Anasarca

Ascites, 10 cc, Turbid Yellow

Pericardial effusion, 36 cc, serous

Edema, 3 + to clavicles

Disseminated intravascular coagulation (clinical)

Hemorrhage, retroperitoneal, small

Petechiae, parietal and visceral pericardium

Hemorrhages, multiple, small (4-5 mm), myocardial at apex

Hemorrhage, mucosal, jejunum, ileum, colon, foci

? Interstitial pneumonitis

Brown pigmentation, pancreas, diffuse

Laryngotracheitis, severe

Mucosal ulceration, larynx and trachea

Hemorrhagic cystitis

Microsomia

Lipid depletion of adrenals

Exhibit 1: Provisional Pathological Diagnosis

Anna Marie (continued from previous page.)

indicated a liver biopsy was necessary which resulted in a positive reading for a diagnosis of *hemochromatosis*.

Irene was 29 years old when she began an aggressive regimen of weekly phlebotomies for approximately 6 months. She continued on an 8-10 week phlebotomy regimen until approximately 2 years ago. For some unknown reason, Irene hasn't had a phlebotomy since then.

Late in December 1978, I reported to Navy "Sick Call" at the U.S. Naval Hospital, Orlando, FL to request blood tests to determine my iron levels. Initially, the doctor felt my request was unwarranted, as I did not have any "overt manifestations." "Your skin doesn't have a yellow cast to it," he said after examining the front and back of my hands. Furthermore, the Navy was experiencing an austere budget, and he felt he wasn't able to justify ordering expensive blood tests. Obviously he needed more evidence, so I suggested he review my sister's autopsy report. Skeptical, he did order the appropriate blood tests.

Several weeks later, I returned for the results. The doctor repeated the results along with the suspected diagnosis several times rapidly. I'm still not sure whether his animated reaction was a result of him being impressed for having seen his first case of *hemochromatosis* or simply being dumbfounded. Regardless, my medical record now reads, "serum iron is elevated to 314...TIBC is 308...and percent saturation is 102%. The patient would appear to have *hemochromatosis*." (A blood test for serum ferritin had not been developed until the mid-eighties.)

The doctor immediately ordered a liver biopsy, along with several other tests. In 1979, a liver biopsy was considered to

be the "gold standard" measurement for a positive diagnosis of *hemochromatosis*. My liver biopsy was performed in January 1979. The pathology report confirmed the clinical diagnosis. "Excessive deposits of hepatocellular iron, consistent with *hemochromatosis*. No evidence of tumor, cirrhosis or granuloma."

In early February, I began an aggressive de-ironing program lasting for 21 months. At first, my phlebotomies were conducted weekly, later they were reduced to biweekly visits. My doctor, David Schneider, Lieutenant, Medical Corps, United States Navy stated goal was to remove the excess iron as rapidly as possible without causing anemia. He primarily used hemoglobin (Hgb) and hematocrit (Hct) as his guide in avoiding anemia since serum ferritin testing was not available at the time.

My phlebotomist, a Navy Corpsman, was excellent at inserting that monstrous 16 gauge needle into my arms! He introduced me to using a blood pressure cuff rather than the standard narrow tourniquet, which allowed me to control the pressure during the phlebotomy and keep my fingers from becoming excessively numb and tingling. He also suggested that I take an aspirin the night before to help alleviate clotting in the needle. I continue to use both of his suggestions to this day, which have made my life-long therapy more bearable.

After retiring from the U.S. Navy, May 1, 1979, my family moved from Florida to Massachusetts in October. By this time, I had accepted without reservation that therapeutic phlebotomies would become an integral part of my life. I first realized the importance of maintaining good records of my medical treatment when I contacted the nearest VA hospital for treatment. I was immediately asked for a copy of my liver biopsy result. I'm glad I had my records, or I might have had to endure the unnecessary risks of another biopsy. Thereafter, I have kept a database of blood tests and phlebotomies, including penetration sites and extraction volume related to my treatment of hemochromatosis.

"The true meaning of her legacy would not be fully appreciated until many years later."

Several years later, I recognized that although Anna Marie's death was akin to one door closing, I had not appreciated enough the fact that my children, and their children, were also impacted by hemochromatosis. I knew it, but at the same time I didn't know it. If it hadn't been for the discovery of the hemochromatosis gene in the mid-nineties, I may have failed to see that a new door of opportunity had opened. Now I could, at the very least, identify which of my children and also their children will potentially be affected. Here was a real opportunity to educate my family. I, along with my sister, Irene, could be the prime example that iron education and treatment would be the key to a healthy future free from iron-related disease for generations to come.

Thus, I embarked on a personal quest to have all my children and their children identified as to the possibility of experiencing iron overload. Genetic testing and establishing baseline values would be the first step and the cheek swab method appeared to be the simplest. My results merely confirmed my diagnosis: a double mutation of Cy282 gene. I did not have my children tested once my wife's results were normal for the Cy282 and H63d genes. Their heterozygosity was a genetic certainty. Using my family as a model, Exhibit 3

Date of Autopsy 9-5-75

Hemochromatosis

Extensive iron deposition in myocardium, liver, (hepatocytes, Kupffer cells, bile duct, epithelium, and connective tissue), pancreas (acini and islets), skin, spleen, gastric mucosa, renal tubular epithelium, lymph nodes, adrenal cortices, thyroid, adenohypophysis, and choroid plexus

Cardiac failure (clinical)

Biventricular dilatation

Pulmonary edema

Passive congestion, liver, slight

Anasarca

Ascites, 10 cc, Turbid Yellow

Pericardial effusion, 36 cc, serous

Edema, 3 + to clavicles

Micronodular cirrhosis, liver, moderate

Fibrosis, pancreas, marked

Hypogonadism

Secondary amenorrhea (by history)

Atrophy, uterus and ovaries

Bronchopneumonia, marked, bilateral (enterobacter cloacae)

Hyaline membranes, lungs

Disseminated intravascular coagulation (clinical)

Fibrin thrombi, lungs

Hemorrhage, retroperitoneal, small

Petechiae, parietal and visceral pericardium

Hemorrhages, multiple, small (4-5 mm), myocardial at apex

Hemorrhage, mucosal, jejunum, ileum, colon, focal

Laryngotracheitis, severe

Mucosal ulceration, larynx and trachea

Microsomia

Lipid depletion of adrenals

Exhibit 2: **Final Pathological Diagnosis**

See **Anna Marie** continued on next page.

Anna Marie continued on previous page.

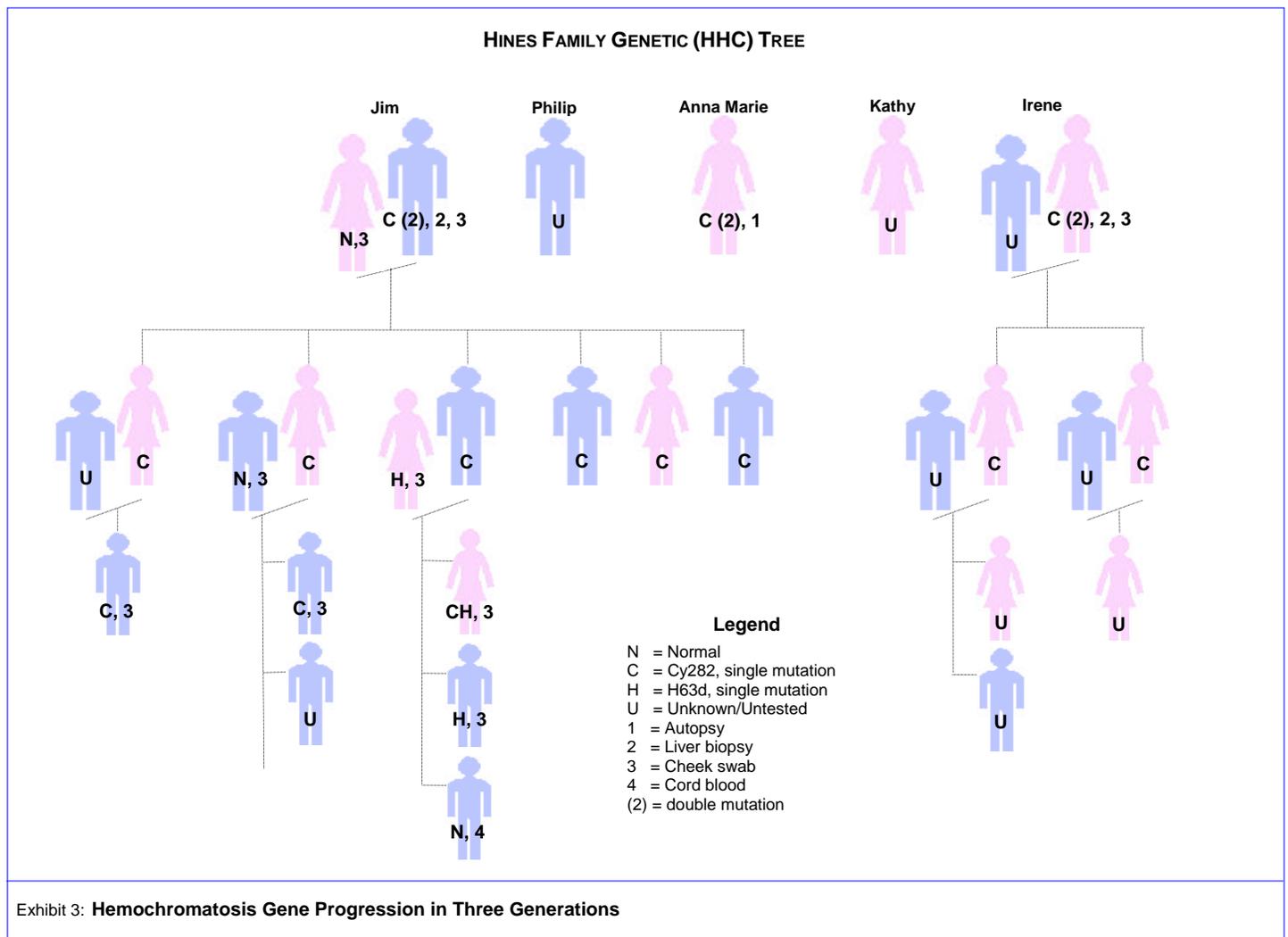
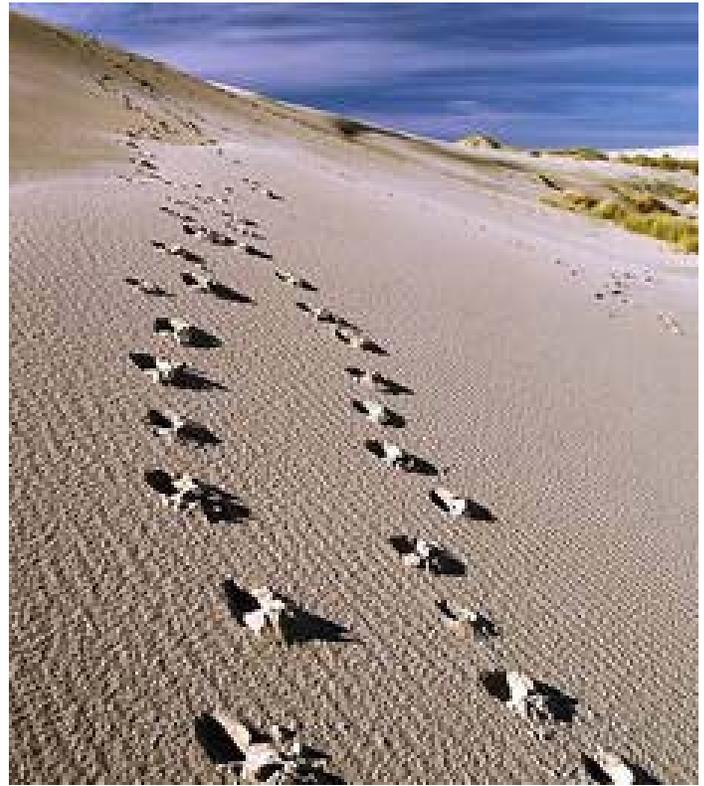
indicates how a specific genetic mutation can show up in subsequent generations.

Additionally, the introduction of a different mutation can complicate the genetic structure.

During the past 26 years, I have enjoyed extremely excellent health notwithstanding periodic phlebotomies to control my iron. My gratitude is perpetual due in large part to Anna Marie's direction, my mother's tenacity and perseverance, and Dr. Schneider for knowing precisely the treatment that was required when I was initially diagnosed. My sister, Irene, is equally as fortunate as I because her doctor was similarly knowledgeable as was mine.

It has been said that our footprints are left in the sands of time when we pass on to the Supreme Commander. If the size of our footprints could be used by future generations to measure our worldly achievements, then surely, Anna Marie's can be viewed as being humongous. In death, she informed her family of what may lie ahead if we don't pursue knowledge to prevent the ravages against the body that can occur from excess iron. Her death also reminds us that it is our responsibility to continuously pass this knowledge on to future generations and to inspire to them to continue to do the same.

That is Anna Marie's legacy!



Dear Loved One,

This letter should have been written long before now as I have always felt I was an important part of your life. On the other-hand you may have simply considered me an obscure, distant kin. Either way, you have known me. At the very least you have known of me. You are no doubt aware of my demise and if our family grapevine is as reliable a source of information as I believe it is, then you are also aware of the suffering that preceded my final and untimely departure.

I may have been your spouse, your parent, your child, or a distant relative. What truly matters is that I was a member of your family, a family with a very strong and vibrant heritage.

Many in our family who preceded you have had the tendency to forget that each of us is a steward of our heritage and have the responsibility to share our heritage with future generations, including the darker aspects. As a living member of our family you are now a trustee responsible to cultivate in the more recent family arrivals our family's ancestral attributes so that the knowledge of our heritage will survive. At the same time you must **NOT** avoid forewarning your offspring of the less than desirable traits. I'm writing to remind you of that responsibility, a responsibility I was extremely complacent about. Had I been otherwise, I might have been able to share this knowledge with you personally.

On the surface, we may not have had anything in common. After all, you and I had chosen completely different vocations, we chose to marry into different families, and we certainly had different opportunities from which to choose. However, we did not choose our parents; consequently there is one commonality we do share, one underlying trait that cannot be denied. Our genes! Yes, I said, our genes. Our genes have been passed to both you and me from previous generations. I must belatedly admit that on average, our family has been extremely fortunate in this regard.

I'm certain you are aware of our family's many obvious genetic characteristics so I won't dally on our family's better qualities. However, I can't impress on you enough that there are also genetic traits that are lurking in the background, which effects may not surface until later in life, and then it may be too late. You have my unrestricted permission to hold me up as a classic example!

Genetic hemochromatosis (HHC) is one of the several genetic traits which our family has inherited, which can result in possible iron overload. Although iron is essential to good health, an excess can be detrimental to your health and just as important, the health of your loved ones. Iron overload can often lead to unnecessary suffering, even premature death, if not diagnosed early and treated in a timely manner. Any one of our family members can be affected because HHC had finally been identified as the root cause of my many medical problems, albeit too late to delay the inevitable. Your watchword must be: **I know this genetic trait exists in one family member; therefore it will exist in others as well.**

You now have the singular opportunity to prevent needless suffering similar to what I had to endure prior to my passing. It's possible that you may also prolong a loved one's life. If you can do that, you deserve to indulge in self-pride. Admittedly,

not everyone who has inherited this particular gene will be affected. Are you willing to take that risk? Consider that my passing could have been delayed for many years had I not abrogated my responsibility and participated more actively in my own health care. One might say that my ambivalence was the cause of my own suffering and subsequent premature death.

For many years, I was reluctant to see my family doctor and had absolutely no idea about my immediate family's medical history. On that rare occasion when I did visit my doctor, I accepted his diagnosis and treatment as gospel and without question. My doctor had been treating me for many health maladies; such as diabetes, arthritis, and heart problems, to mention a few. I was convinced, because he had always been my family doctor, that I was receiving the proper diagnosis and treatment. I trusted him.

Allow me to digress for a moment to relate a little story, which may emphasize my point more cogently.

Several years ago, my wife could not start her car when she left work. The engine would not turn over when she turned the ignition on. I was out of town, so she had her car towed to our mechanics shop, a trusted mechanic because of previously good maintenance on her car and because she had little or no knowledge about an automobile other than to drive one. She went home while the mechanic went about diagnosing the problem. The next morning, my wife went to retrieve her car with a newly installed battery assured that her car would start. However, the engine would not turn over. So she returned home. The following morning, she received another phone call that her car was ready and able to start. My wife returned to the garage once again where for the third time, she was unable to start the engine. Finally, the mechanic decided to look beyond his initial assumption that the problem was a weak or dead battery. What he found was that an electrical switch in the car's air conditioning unit was shorted out causing the battery to lose all its power in a relatively short time. Once the switch was replaced, my wife had no more problems starting her car.

Why did I pass that tale on to you, you ask?

Well, my wife's battery experience was a classic example of treating symptoms, rather than seeking and fixing the underlying problem that created the symptoms. Fortunately, her experience only resulted in several dead batteries. Not, as in my case, an early sentence to eternity.

My medical problems were not improving under the care of my trusted family doctor, so I sought a 2nd and a 3rd opinion. Finally, on a visit to the 4th doctor, I was diagnosed with genetic hemochromatosis. My symptoms; arthritis, diabetes, heart problems, etc. were being treated rather than the cause of those symptoms. Had the actual cause, an excess of stored iron, been identified and treated earlier I may not have had as many medical problems. Alas, the damage from the excess iron to my various organs was irreversible. Well, you know the rest. What is even more ironic, had I been just slightly altruistic and donated blood, and done so frequently, I

would have forestalled the unnecessary suffering I have mentioned. Of course, hindsight is usually 100% correct.

It must be clear to you by now that decisive action is essential to protect the future health of your loved ones. Obviously, it is your choice to pursue the requisite knowledge. To do nothing is tantamount to sentencing someone in our family to a quality of life that may include serious medical problems, even death. Take, for example, my eternal status.

You have an edge that wasn't available to my generation. The internet, for example, allows you access to a cornucopia of information that can only increase your knowledge of the effects of iron overload and to afford you the opportunity to actively and intelligently participate in the decisions that impact you and our family's health. All this can be accomplished within a relatively short period of time! Please take advantage of these fantastic educational tools. They can and will provide you with a formidable knowledge base with respect to stored body iron levels whether it is in excess or in short supply.

Not only is the internet available to you, there are also a variety of support groups, both on-line and locally, whose members willingly offer to share their personal experiences in how they have dealt with excessive stored iron in their bodies. These wonderful people share their experience and acquired knowledge with the hope that you do not have to endure similar frustrations they had experienced in their quest for a proper diagnosis and treatment. These remarkable human beings have formed congenial and knowledgeable support communities. You are freely welcome to become a member of their communities.

You may have noticed that I have not mentioned any of the details about the diagnosis or treatment of hemochromatosis. I purposely avoided sharing the details with you as my intention is to impress on you the importance of the message, iron can kill if left to accumulate to unmanageable levels. Your inquisitiveness will be sufficient to find the details. The specifics are abundantly available to you and every living member of our family who are willing to seek this knowledge. You will discover that both the diagnosis and treatment are relatively inexpensive compared to the alternative of doing nothing. Although treatment will be a life-long journey, you will readily recognize that it is simply plain old common sense. Please keep that in mind.

In closing, I beseech you to be in the vanguard for our family. Be the standard bearer for the continuity of our heritage. Persevere when you are frustrated in your attempts to share your newly acquired wisdom. You will encounter considerable skepticism and denial. Recognize that you can lead a horse to water, but you cannot force them to drink. Nevertheless, be prepared and keep available the water at all times. I can assure you from my vantage your efforts will be well rewarded.

I will continue to remain eternally

Your loving family member

THE RUSTY CURMUDGEON

Jim Hines was diagnosed with hemochromatosis in 1978. He is living testimony that an early diagnosis and participatory iron management will provide longevity along with improved quality of life. Jim can be contacted at jedwhines@cox.net



"If you have knowledge, let others light their candles with it."

Winston Churchill, *British statesman*

PORCUPINES AND HEMOCHROMATOSIS

Most hemochromatosis patients encounter "porcupines" almost immediately after being diagnosed and after the realization sets in as to how detrimental hemochromatosis can be to their family and subsequent generations. Sort of like an epiphany. Consequently, we singularly assume the responsibility of attempting to warn as many family members as possible of what action should be taken to avoid the potentially ravaging effects excess iron can have on our bodies.

However, we quickly discover that our new found knowledge, not too mention our sincerity is met with indifference and ambivalence, and in some cases out and out hostility. I know you've encountered this attitude. I've been there, done that! It's hard to miss this attitude when the individual you are trying to educate rolls their eyes until all you can see is white, or they tune you out by closing those invisible ear valves while looking you straight in the eye, not hearing a word you are saying. It's really frustrating when you know in your heart and believe that you are actually saving them from potentially serious and long-term morbidity. Possibly death! You can almost hear their thoughts, "Here he goes again, spouting all that stuff about iron." "Do I have to hear this again?" I hate being "dissed." I actually had a cousin, who happens to be a nurse, inform me that that hemochromatosis couldn't possible exist in her side of the family. Many of you have probably had similar experiences.

In these situations, we have a classic example of the "Porcupine Syndrome". Porcupines move close to each other for warmth, but then get stuck by each other's quills, then they move apart, but they get cold and so move close and back and so on and on. You see similar behavior every time we HHC patients broach the subject of iron, our family members try to avoid us and the topic, much like avoiding quills. When we shift subjects, they once again embrace our kinship and warm to our relationship. Dr. Margaret Mahler, an eminent child psychologist, refers to this behavior as "ambitendency," a tendency to contradictory behavior arising from conflicting impulses.

Having frequently experienced this dilemma over the past 28 years, I knew some attention-getting method had to be devised; obviously talking was not working in many instances. I just never knew what that something could be. It wasn't until Patrick McKeever submitted his poetry (See page 2) for publication in the newsletter about this HHC community problem did I realize that a letter from a person who died from hemochromatosis complications to living family members might just work. I also observed that describing blood tests, spouting blood test ranges and values, and encouraging DNA testing also turned many people off faster still. Thus, I decided to write "A Letter from the Afterlife" (See pages 9 – 10) devoid of these characteristics by appealing to their integrity and curiosity by encouraging letter recipients to act responsibly and consider the well-being of their loved ones.

You are welcome to use this letter in its entirety by simply printing only pages 9, 10 and 11 of this newsletter. Or, if you would like to modify the letter, for example, addressing the letter to a specific family member you are welcome to do that also. IDI has provided a text file, which can be viewed using the link at the bottom of this page.

This page will open in the Windows default program, Notepad, you simply have to save it, or copy it to your favorite word processing program.

Hopefully, "A Letter from the Afterlife" and Patrick McKeever's poem, *I am a Famous Person Who Died of HH*, will be valuable tools in informing your loved ones about the dangers of iron overload.

Searching for the ideal holiday gift for a loved one!

*Visit our **Iron Store***

<http://www.irondisorders.org/Store/>

Menopause (Continued from page 1.)

women are likely candidates for recommendations of supplemental iron, which may be unnecessary. Fatigue is symptom of iron deficiency, but fatigue is also a symptom of anemia of disease or deficiencies in B12, folate or zinc.

Anemia of Chronic Disease

In adult females, anemia of chronic disease is likely due to some common ailment such as urinary tract infection, a head or chest cold, mononucleosis, tonsillitis or strep, stomach or intestinal flu, and bacterial infections such as *H. pylori*; in some rare cases tuberculosis. Most of these conditions are treatable and when the patient is cured, the anemia will be corrected. If the anemia persists once an illness is corrected, the doctor will want to investigate further for a secondary underlying cause of anemia that may be more serious such as lupus, thyroid disease, or cancer.

Preventive Measures

To keep iron in balance, ask for a complete iron panel at the annual check-up. Other measures can include: Drinking tea with meals; tea contains tannin that impair the absorption of iron

Donate blood regularly. Limit supplemental vitamin C to less than 200 milligrams per dose. Stop smoking and cut back on alcohol consumption.

To learn more about iron overload, anemia of chronic disease and iron deficiency anemia visit our website.

"A Letter from the Afterlife" text version – <http://www.irondisorders.org/Newsletter/AfterLifeLetter.txt>