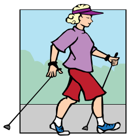

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ECD Global Alliance News

ECD

By Dan Perch

I am writing this article directly to those individuals afflicted with Erdheim-Chester disease (ECD), and their families and caregivers. While those of us diagnosed with ECD are few in number, I believe that ECD is an extremely under-diagnosed, and therefore, an under-reported disease. My conclusions are base on my own research of many of the 300+ printed studies on ECD[1] and the few personal interviews with other ECD patients and caregivers. This disease affects each of us in different ways. I want to share my experience, and at the same time, provide some hope and encouragement to you as you face your personal challenges with this disease.

I am 63 years old. After 20 years as a military intelligence officer, I retired from the US Army in 1991. I then began my "second career" as a cyber security specialist for a Federal government agency – nearly 18 years now. I plan to retire in three to four years – from the government – but not life itself. Recently, I have been reflecting on what I will do after retirement. These two careers might seem quite divergent. Twenty years ago, they were. I had to "re-invent" myself and make the transition when there were no standards for IT security or for the IT security specialist. Cyber security was a term not even invented! There were only "best business practices." I pursued each vocation with a passion. It was more than a job. I enjoyed the work. I loved it. Each vocation had its set of responsibilities and challenges - constant, but changing ones. I still wake up wondering and anticipating what challenge is going to greet me that day.

When I am not "working", I have other interests, hobbies, and activities; too numerous to list, but the one I am most passionate about is sports. I have actively participated in sports through out my life. In high school, it was football; in college, intramural sports. During my 20 years on active duty (and in addition to the required semi-annual US Army physical fitness test[2]), I participated in flag football, basketball, softball, bowling and one year (at age 42), as a player and defensive line coach for a semi-professional German-American football team in Europe. My sports regimen since military service has been self-limited to co-ed league softball – with the 20- and 30-somethings, as a player and coach.

I want to spend more time with my two adult sons, their respective spouses, a two-year old grandson, and my fiancée, Rita. Rita and I plan to marry soon. However, all these plans and expectations received a jolt three years ago. I paid a visit to my internist, complaining of slight edema in one of my ankles. My complaint was a symptom remaining from an ankle injury endured three or four summers ago playing league softball. I simply twisted my ankle, stepping down on base and catching only the base's edge with my cleats.[3] It took forever to heal and the "black and blue" marks never entirely cleared up. Several tests from the customary clinical laboratory tests to Doppler and CT scans were ordered. My internist wanted to eliminate the possibility of clots, blockages or other restrictions of the arteries in my leg. She called me the following day informing me the results were in and she needed to see me "right away." The lab results were normal; the CT scans results were not. They revealed each kidney

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"Simple Simon Says…"

Planning Your Day

Planning your day is an important thing to do. I think that this is especially true for those of us who have led a very, regulated life. This might have been related to a job, or mainly home and family based. I was a family physician when I last worked, and this was very organized. I had a strict weekly time-table. I could predict what I would be doing on, say, Tuesday Afternoon on the 14th of January next! Going from this degree of organization, to a routine where you have virtually no fixed activities, is not an easy thing to do. At that time, I didn't need to "Plan my day". It was already "Planned" for me.

I have heard of a retired man who plans his life on a spreadsheet! This is too fancy for me! I have a pile of blank paper and a pen on the Breakfast table, and I write on this what I need, or want to do. I also am fairly hooked on my diary too. I need it for all future planned events.

I'm sure that it is therapeutic (doctor-speak for "good for you") to make a plan with specific items on it. It can help you to organize your day so that you do achieve some things. And, you are less likely to forget to do some of the regular things (making the bed, filling or emptying the dishwasher) that are appreciated by the rest of your family when they get home at the end of the day.

You can look at your list, and it helps to make you realize that you *have* done something with your time. Your family, have the opportunity to keep an "overview" on your activities, so that you aren't taking on things that are beyond your capabilities. Also, there probably, won't be any suspicion that you have been sitting around doing nothing. Exactly what you choose to be in your plan, will depend on how your illness affects you. Some of us will be able to do quite a lot more than some others. I think that it helps us to maintain some level of self-respect, if we can contribute in some way, no matter how small. I was used to full-time work with a good income. I felt that I could justify this by the amount of time and effort that I put into my work. Things have now been completely turned on their head. Planning the day helps me to cope with this massive change.

Yours,

Simple Simon

~You gotta love living, kid,
cos dying's a pain in the ass.

About the ECD Global Alliance

~New Information~

www.erdheim-chester.org
support@erdheim-chester.org

The ECD Global Alliance has made the first steps toward becoming a non-profit organization, also known as a 501(c)3 organization. We are officially a corporation with a formal Board of Directors. It will take several months, but once established as a non-profit organization we will be able to receive tax deductible donations. It is hoped that enough money can be raised to support ECD-related research directly. Please continue to check the website www.erdheim-chester.org for the most current updates on this status.

If you would like to make a donation or get involved in any way, please email the ECD Board of Directors at support@erdheim-chester.org. Together we can make a difference!

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encased in a large tissue mass! There were no outward physical signs or other biological indications I had this abnormality. I was in excellent health. A preliminary diagnosis of retroperitoneal fibrosis was rendered based on the CT scan. More tests would be necessary to be the diagnosis definitively. Were it not for the ankle complaint and my internist, who is meticulous at getting to the root of things, we still might not know anything was "wrong."

Throughout my entire life, I have been reasonably healthy: the usual childhood diseases and the seasonal cold or flu, but no broken bones or serious injuries. Whenever I have taken ill, my recovery was normal within the expected timeframe.[4] Medical help was always available, usually efficient, and proper in diagnosis. The first inclination that I was facing something serious was the length of time and the number of well known, respected specialists stumped by my newly discovered ailment. They consisted of a hematologist, an oncologist, urologist, and a radiologist. Things they did agree on: It was not cancerous, melanoma, Hodgkin's or non-Hodgkin's. It was neither contagious nor genetic. I was told everything I did not have versus what I had. It was idiopathic![5]

An initial diagnosis of retroperitoneal fibrosis - 10 months of more testing, and two biopsies - proved to be inconclusive – twice. The surgeon performing the second biopsy, after some study and research, referred my case to a nephrologist at Johns Hopkins University Hospital (Baltimore). Now, I received a preliminary diagnosis of Erdheim Chester Disease. A bone scan would solidify this diagnosis. (For those not familiar with the bone scan, the patient receives, intravenously, a small amount of a radioactive isotope. Thirty minutes after injection, an x-ray scan of the entire skeletal is performed. If ECD is present it will be pronounced in the long bones of the lower legs and forearms. As my doctor stated, "If you have it, you will light up like a Christmas tree on the film". Well, I lit up!)

For the first time in my life, I am confronted by a most bizarre medical situation: dealing with a disease whose cause is unknown, is random and inconsistent, can strike other body organs beyond the long bones and for which there is no known or proven cure. The feeling I experienced was like being adrift in a boat at sea (as in ocean). There is no land in sight during the day. At night, the sky was clear, filled with plenty of stars, but, they are of no use. I have no map; and even if I did, it would be of no value since the stars are all foreign, unmapped, and I have no reference point.

While the doctor had prepared a "maintenance plan" for me, I wanted to get a "second opinion" – not because I did not believe him or his diagnosis, but because I had already been through one "mis-diagnosis". The literature on the drug he was recommending stated that once you start it, you should not cease taking it without further medical evaluation. (Apparently, whatever one is afflicted with could come back more aggressively.) Notice, I said 'maintenance plan'. How can you 'treat', let alone 'cure' a disease of which its genesis or cause is unknown? Nevertheless, I asked if I had to make any dietary changes, life style or any other activity changes in life. There was none. I became firmly resolved that I was not going to change the way I lived in manner or style. I would remain actively engaged in all my activities: work, sports, social, and family. I was not going to let ECD consume my life. Furthermore, I was resolved to learn as much as I could about this disease and beat it.

Now, my fiancée, Rita, would say I am a pessimist – always taking the worse case scenario. I am always countering (as in defending);"I am a realist." She has been a rock of support throughout this challenging time. She was there when I receive the preliminary diagnosis of ECD. She traveled with me to the Mayo Clinic (Jacksonville, FL) for the confirming second opinion. She has been there with me for each of my semi-annual, follow-up appointments with my ECD physician at Johns Hopkins.

I will continue to do what I can do for my loved ones and myself. I know from experience sometimes you have to turn things over to a Higher Authority! This is one of them. My faith has been my main rock.

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So my words of encouragement:

Remain active in life – in mind and body. If you are not, become active. Within your limits (and after checking with your medical professional), become involved in an activity or interest that you love to do or are "passionate". Pick it up. Pursue it! I know some of you are physically tired or frequently fatigued (from ECD and/or effects of medication you may be taking). Can you walk? Do you know how far you can walk before you tire? Can you add a little distance each time you engage in this or other physical activity?

Take charge of your health care as you face each challenge. You are your own best advocate. ECD Global Alliance is a great resource. While they specifically do not render any medical advice, there are more medical professionals known to it who are researching ECD and/or treating ECD patients than a year or two ago.

Emotional support of your family, extended family, and close friends is also important. If you lack or have none, contact the ECD Global Alliance. They can put you in touch with me. I will listen.

Dan Perch

Footnotes:

[1] Erdheim-Chester disease was first discovered and reported in 1930. The 300+ case studies I am referring to are those found in National Institutes of Health's PubMed Library and only date back to October 1988.

[2] For those unfamiliar with the mandatory, semi-annual US Army Physical Fitness test (of the 1970s and 1980s), every member had to complete a standardized number of repetitions [based on age] of pushups, sit ups, chin ups, "walking" the monkey bars and complete a two-mile run – in combat boots – at or under a prescribed time. We also had to meet weight and body fat standards based on height.

[3] I was safe!

[4] It helps too when your mother is a Registered Nurse: US Army nurse in World War II (North Africa and Anzio, Italy); mother of five children; a masters degree in Nursing; and taught Obstetrics for over two decades at the local hospital.

[5] They must teach this definition to first year medical students: an idiopathic disease is one which the cause of, when and where in the human body it occurs, and the cure for it – are all unknown to the medical profession – making the physician appear to be an idiot. I have had three different physicians/specialists inform me in this fashion.

Online Chat Information

http://www.chatzy.com/427437211406

The ECD Global Alliance hosts scheduled online chats each week. Anyone affected by ECD is welcome to attend. This includes patients, caregivers/carers, family members, medical personnel, etc.

For those who register with the ECD Global Alliance, emails are sent out announcing each chat session and a summary email is sent following each chat session. The time for the next chat session is also always posted in the chat room. If you have not yet registered with the group, you can always enter the chat room to find the time of the next scheduled chat session.

The chats are open for any discussion topic within reason. Often discussed topics include: ECD diagnosis issues, treatments, tests, knowledgeable doctors, symptoms, treatment side effects and sometimes just friendly discussion about other pleasant things happening in participants' lives. Most who attend the sessions have found it a good way to meet others whom they can correspond with outside of the chat sessions. For some this has proven to be a good way to build a support structure of others who are interested in learning as much as they can about ECD and providing mutual support to each other.

We invite anyone affected by ECD to attend and participate in the chat sessions.

Letter from the Editor

As we complete our fourth edition of the ECD News, I would like to take this time to introduce myself. My name is RuthAnn Batchelder (screen name Yankees Fan). I have been involved with the ECD Global Alliance since before it began. Carol Smith, Kathy Brewer, and I began communicating in 2007 about how we could help others affected by ECD. After much discussion we held our first online chat session in April 2008. As the months passed I was unable to dedicate as much time as I would like, so as of today my remaining contribution to the ECD Global Alliance is as editor and publisher of the newsletter.

My husband, Lance, was diagnosed with ECD in 2006 after his initial diagnosis of Langerhans Cell Histiocytosis (LCH) in 1995 – just 15 months after we were married. As Lance's health declined we were forced to open up more and more about the effect it was having on our life. One day we received an email from a family member stating that it was our own fault that Lance was sick. It was a ridiculous statement, and I knew it, but the fact that a family member could be so hurtful was beyond my comprehension. I have always been a very private person, and it had been extremely difficult for me to open up and let our friends and family know what was going on in our life. At that point I started to shut down, not allowing people to be a part of our life and not allowing them to help us anymore.

I know we cannot let one statement ruin our life, but it is eye-opening how a little bit of negativity can have such an impact on one's life, especially during times of great stress. Lance and I wouldn't make it through our days without our faith, or all the support we have from the rest of our family and friends. We have started sharing our life with others again…now with more caution…and a new plan. To overcome any negativity in our life we find something to be thankful for. It is amazing how different life looks when viewed with a positive, thankful attitude instead of allowing the negativity to take hold. Don't get me wrong – it can seem an insurmountable task to find something to be thankful for in the midst of turmoil or trial. But the exhilarating feeling of triumph over negativity cannot be matched! I urge you all to give it a try. You will not be disappointed!

The ECD News was started as a way to support each other and share our stories. We need to hear from you! If you are living with ECD or caring for someone with ECD (or are a friend, relative, or medical professional) and would like to share your story or what you are thankful for, offer support, advice, or words of encouragement, please email me at **stanceforlance@hotmail.com**. *If you would like to submit your story but need help writing or editing I am available and happy to assist.*

A big thank you to all the volunteers of the ECD Global Alliance, the contributors of the ECD News, the medical professionals working to make our lives better, and the countless friends and family who support all who are affected by ECD. Together we are making a difference!