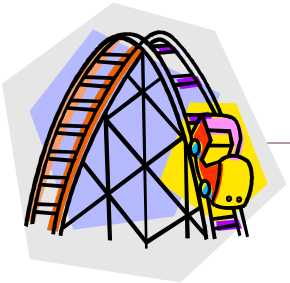


Issue I

January 2009

ECD GLOBAL ALLIANCE NEWS

ECD Global Alliance News

Sharing Our Stories:

"Living with a Rare Disease – One Couple's Perspective" by Carol

And then we finally knew. We knew that our lives together would be altered from that moment forward. It was a great relief to finally know the reasons behind all the illness, but on the other hand it was not what we had hoped ....."you have a very rare disease – it is a progressive disease and we do not have a cure for it and further we only have experimental drugs to try to treat it".

Now nearly three years later I can still hear those words being spoken to us. I can still feel the chill that came over me as the words were spoken. And yet, I still cannot believe this has happened to us. Us – are we so different than many other couples that have had similar words spoken or who have faced tragedy? No, we are just like all the rest when those words are spoken – we were scared and didn't know in which direction to turn. We drove home that day from the hospital in silence – silence of disbelief.

What did those words mean? They meant that WE have a rare incurable disease that nobody knows anything about. The disease is Erdheim-Chester Disease (ECD) a non-Langerhans multi-system histiocytosis of unknown cause. At the time of diagnosis we were told that ECD had already been seen in the long bones and the bone marrow as well as the lower spine, the kidneys and the abdominal aorta. We were told there was an experimental drug therapy that had been having good success and that it would be started as soon as possible. "We hope it will slow the progression of this disease down, and by doing so it may prolong your life" the doctor said.

What has happened in the past two and three-quarter years? We started the drug therapy and so far it has been successful. It has indeed slowed the disease progression down. We have come back from the brink of death and now live a relatively normal, albeit greatly altered life.

Some days following his shot (a self injected shot 3 times per week) Mike feels good and other days he has fever, chills and is really weak. He takes at least one nap a day and tires very easily. Living with any illness is difficult all by itself, but then you add in RARE and you find that your options are very slim. Not only are there no doctors willing to treat (they maintain) but there is no information available to help you understand. There are NO answers, only more questions. Rare diseases are referred to as orphans, when in fact the real orphan is the one with the disease. It is like taking your car in for repair to your local hospital. Or like going to a mechanic when you are ill – same difference as neither can give you any answers or help you fix the problem.

Every day is a new challenge either with the one affected by the disease itself or the spouse or loved one who has to deal with all the other daily stresses such as health insurance or the lack thereof, inability to earn a living, not to mention how to pay for such an expensive drug?

I have heard many people say "you never know what the future has in store" and "you have to be ready for whatever may come along". Now we know, but we also know that you can never be prepared for a rare disease diagnosis.

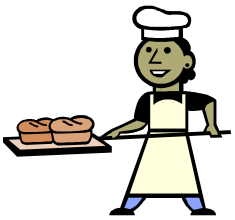
Our lives have changed mostly in our daily routines. Our outings consist of trips to the grocery or drug store and every three months (down from weekly and monthly) to the doctor. We don't go out to dinner any longer, or take vacations as we can no longer afford such luxuries, but we enjoy the simple pleasures of being together, watching television and feeding and caring for our dogs. The dogs you say? There was a time when Michael didn't have the capacity to care or love an animal, but now the dogs seem to be what he cares about most (and me too).

We have surely learned a lot over the past two plus years not the least of which is how much we love each other and how very precious our time together is. It seems that there is always a silver lining to be found.....even in the rarest of places.

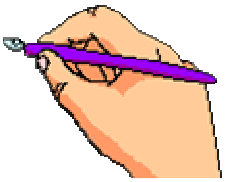
Caregiver to Caregiver

"What About Me? Avoiding Burnout." by RuthAnn

We all seem to be at different stages as caregivers, with some of our loved ones requiring more care than others. But one thing that is true for all caregivers/carers is that we need to take care of ourselves so that we can better take care of our loved ones. This is something I learned very early into Lance's illness, and I am so thankful that I did!



It may not seem to someone on the outside that things are all that difficult for me, after all I am not the one who is sick. But I have had to learn to live with all the changes from this disease, just as Lance has.



We were in our early 20's when we married. Our whole life was ahead of us. We had so many hopes and dreams. This was the man I was going to spend the rest of my life with—my partner, my equal, my best friend.

Lance was diagnosed with LCH 15 months after we were married (we found out later that it was actually ECD). When you marry someone with the intention of spending the rest of your life with them, you figure that after retirement age one of you may have to take care of the other. It is not something you plan to happen in your 30's.

When he was first diagnosed we knew that our life would never be the same, but we had no idea what the future held for us. Thankfully, we both had the same idea—we needed to live life and take advantage of every possible opportunity to enjoy our time together. We traveled, went camping and fishing, attended baseball games (YankeesFan), spent time with friends, etc.

During the early years of Lance's illness doctor appointments were less frequent—about every 3 or 4 months—unless something more serious came up (ie: surgery or radiation). During the more trying times we would buckle down, get serious, and get through the tough times. And after that I would always make time for myself (shopping was first on my list).

Today I look back on those years and I am so thankful for all the opportunities we seized when we had the chance. Two years ago Lance's disease had progressed to the point where he was no longer able to work, and he retired at the very young age of 37. And just a few months ago I made the difficult decision to quit my job so I could be at home with him.

This is not something that either one of us asked for, but here it is and we have no choice but to deal with it. There is no denial when you live with a disease 24 hours a day, 7 days a week. It is like having an unwelcome third person in the marriage.

Since there is no known cause or cure, and no guaranteed results from treatment, we have absolutely no control.

So what does a caregiver do in what seems like such a dire situation? I figure I have two choices every day when I get up: I can either let this eat me alive, OR I can be thankful that I have one more day with the man I love. Most days I choose the latter, but everyone has their bad days right?

But being thankful and trying to be positive are just not enough. There is so much that goes into caregiving—physically and emotionally. As caregivers we need to make sure we are taking good care of ourselves. We need to recognize that our needs are just as important as those of our loved ones, otherwise we get burned out and will not be at our best when we are really needed.

Because Lance and I are on a very tight budget these days I have had to be creative when looking for ways to spend my time. It has been a bit of a challenge to find things to do that require little money. Here are just a few ideas:

~Go through your house. I bet you will find all kinds of things you can do, projects that have never been finished (or maybe never even started).

~Find a quiet spot and read a book.

~Spend some time in the kitchen cooking or baking.

~Write a letter to someone you love. It is so nice to actually receive something in the mail instead of via email!

~Get out of the house and go visit a friend. If your loved one is unable to stay alone ask a friend or neighbor to sit with them so you can have some time to yourself.

~Turn on some music and dance, or just sit and enjoy.

~Exercise!

~So you can't go have that expensive coffee, but you can make coffee at home and invite a friend over—or call someone long distance and have coffee over the phone. (I do this with my mom at least once a week.)

~Go to a quiet room and meditate.

~Put together a puzzle.

~Write in a journal.

~Call a friend and connect or reconnect. Do not lose track of those who are important to you. And know that it is okay to talk about your feelings and your fears!

Please remember to take care of you and learn to recognize when you need your space. We do not spend time away from them because we do not love them—*it is because we love them so much*!

"Simple Simon Says…"

Hello All, and Welcome to the newsletter.

I have been asked to write some pieces for you. I intend these pieces to be informative, helpful, and, hopefully, entertaining. Much of what we, and our loved ones, have to go through is not any of these things, and I hope to redress the balance a bit. If I sometimes seem to be a bit silly then I'm sorry. I don't intend to offend anyone. If the columns don't live up to expectations, then they will be stopped.

I was a family physician for about 20 years in the North of England, before I became ill. I lived an active life, both at work, and also at home (my wife and I had a standard *(large and not fat!)* poodle dog. 63 pounds of stupidity and muscle!). These pieces will, naturally, have a UK perspective but I will attempt to adapt them to make them helpful to those in other parts of the world.

I don't want these pieces to appear like opportunities for me to brag about what I'm up to. I don't want them to end up being "ME, ME, ME".I would like people to think about what *might* be possible, and how to go about it safely. For instance, some ECD patients have found that a tricycle is suitable for them for exercise and transport. I *did* try using such a machine, but I found that my balance was not really good enough for safe use. So it was *not* for me, but it *may* be for you. We need to try these things out to see whether they are useful to us.

I should now apologize to any of you who have ploughed through the next bit in another piece. I'm afraid that I think that it is worth giving an account of how I got to my present position. I think that by comparing our individual routes to diagnosis and treatment, we can often learn things.

I got my initial diagnosis of ECD about 4 years ago. I had been having double vision and went to see the eye surgeon who had fixed the same problem a few years before. He asked me how I was generally, and I replied that I was a bit wobbly on my feet and slurry of speech when I walked in the woods with my dog. He ordered an MRI scan. This showed tissue behind my eyeballs (retro-orbital). Nobody knew what this was and I went to see an eye surgeon in Leeds Teaching Hospital. He didn't know what it was either, and was seriously considering whether to take a biopsy to find out what it was. Before this happened, I had a generalized seizure and fell into the hands of the local neurologist. He put it all together and came up with a working diagnosis of ECD. I then was sent to London to see a specialist, who ran a unit for another form of histiocytosis (Langerhans Cell Histiocytosis). He started off the clinic visit with, "I've heard of this disease; you've got it; I've *never seen* a case!" He started me on chemotherapy and interferon injections.

He started off the clinic visit with, "I've heard of this disease; you've got it; I've *never seen* a case!"

Initially this was azathioprine, which upset my liver, and then cyclophosphamide. While on the cyclophosphamide I had a bad chest and was sent off to hospital. I was an in-patient from the end of August 2007 until early November! A couple of weeks were spent in Intensive Care on a ventilator. My wife was told on more than one occasion that I only had a 5% chance of surviving through that night. Luckily I have mainly recovered from all that excitement, but I don't want to repeat it.

After I came home cladiribine therapy was suggested. We did not want to have this treatment supervised by a hospital in London. There is a Teaching Hospital in Leeds, just a cab ride away. I contacted the Professor of Haematology there, and I have had cladiribine therapy over the last 6 months. I am now awaiting some more scans to assess whether the cladiribine has helped. I am definitely able to do a bit more than I could!

This all means that I have been THERE and I bought the T-shirt! Perhaps because I have been on both sides of the fence, I may have some insight into the overall situation. At least I knew, personally, a number of the staff involved in my care before I was ill. I am sure that this makes the atmosphere a bit more relaxed.

So, that is my story, I shan't inflict it on you again. Hopefully I can be more entertaining next time!

Yours,

Simple Simon

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

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

About the ECD Global Alliance

For those of you living with or caring for someone with Erdheim Chester Disease (ECD), you know how frustrating it can be to find answers and support for the daily struggles faced with such a rare disease.

In Spring 2008, a small number of ECD patients and their loved ones decided to form an unofficial group with the intention of providing support, raising awareness, and promoting education of ECD.

Thanks to our 100% volunteer effort we have accomplished many things in just a few short months: created a website devoted to ECD; established online chat room and continue to hold weekly chat sessions; facilitated a teleconference between Dr. Kurzrock, an ECD knowledgeable doctor, and patients; found a doctor (Dr. Haroche) doing research in ECD and made that information available to patients; published a patient log that summarizes numerous ECD cases; published a brochure on ECD; provided the first personal and patient-to-patient support structure known for ECD patients; and we continue to do more!

Global Rare Disease Day Set for February 28th

The ECD Global Alliance will participate in a global Rare Disease Day on February 28, 2009. As a Rare Disease Day Partner we will join hundreds of other patient organizations, government agencies, medical societies and companies in focusing attention on rare diseases on that day.

The theme is that rare diseases are a public health issue, affecting millions of people around the world. The hope is that Rare Disease Day will increase awareness of rare diseases, the special challenges encountered by those affected, and the need for research to develop safe, effective treatments or cures.

Please visit the website at http://www.rarediseases.org/rare_disease_day/rare_disease_day_info for more information and to download a sample letter to send to your governor requesting that February 28th be declared rare disease day for your state (USA). Visit www.rarediseaseday.org for the global website.

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.

If you have any comments or would like to submit an article for a future newsletter please email the editor at stanceforlance@hotmail.com. Feedback and input is always welcome. Together we can make a difference!