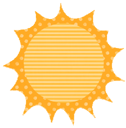

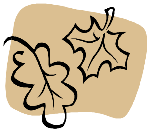

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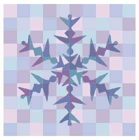

ECD Global Alliance News

ECD GLOBAL ALLIANCE NEWS

Erdheim-Chester Downunder

by Charles Balnaves

It was something of a shock to find some years ago that a simple deterioration of vision, fluttering in the peripheral vision of both eyes, did not imply the need for my wife to wear glasses, a hurdle I had crossed a year or two earlier with no problems other than the need to learn new habits (spectacles don't like being sat-upon – they break and sharp shards stick into sensitive areas). No, this 'fluttering' required a biopsy from behind one eye to attempt (partially successfully) to diagnose the cause of the growths around the optic nerves causing this change in Tanya's vision.

Two years earlier I was sitting in my hotel in Houston (Texas, USA) 15,000 kilometres and days travel from my normal abode in Melbourne Australia, when my phone rang and my son informed me that he had taken Tanya to hospital for a routine angiogram (of which I was aware) and she been given an emergency stent, inserted into the right coronary artery, because she had in fact been just two days from a massive heart attack. But!, says my son, she's perfectly OK and comes out of Hospital in two days! It was a long trip home.

For an engineering manager with global responsibilities, flying around the world and away from home for much of the year it was time to take stock and though I might not have phrased it quite so at the time, a verse from Ecclesiastes was relevant: "To everything there is a season, and a time to every purpose under the heaven" (Ecc 3:1). I had been doing the global travel thing for a decade in one form or another and perhaps it was time to make a change.

The thought went no further then, other than to make sure that my schedule enabled me to be in Melbourne for the biopsy and immediate following period and to be around for most (not all) doctors visits after. It was two years more before I stopped flying around the world and then the change was due to (involuntary) redundancy (a marvellous opportunity to start a new, Melbourne based, but completely un-known and un-anticipated vocation).

The biopsy started a process which led to diagnosis of Erdheim-Chester Disease over five years later and ongoing treatment controlling the disease up to the present. It is a process in which all the specialists did an excellent job while struggling to gain understanding of something so rare not one of them had even heard of the disease. It is also a process which has led to some difficult changes and some wonderful revelations for myself and for Tanya.

Tanya took early action, lots of credit to her there, and that's a first message from us: don't let odd changes go unexplained, go to the doctor! Early diagnosis saved her eyes and minimised the later downsides of Erdheim-Chester Disease: for her and for us. This early action and the excellent onward-referral to appropriate specialists led to treatment of Tanya's case of Erdheim-Chester Disease in a way which means she is not debilitated. We have none of the stories of brushes with death. Rather we have an evolution of life.

Tanya has had to learn to manage her time carefully as she gets very tired very easily. She's had to learn to manage continual significant pain and we've both had to learn to be careful where she goes and who comes to visit ("If you have even a 'little' cold, please don't come visiting" is a mantra I have had great difficulty coming to terms with but it is essential as a 'little' cold for me is months of serious illness for Tanya.)

I've had to learn to be available when needed – not as a 24/7 carer, but to help ensure that things get done, to take on some of the household duties as 'my' tasks, to help with others as needed, to go along on (most) doctors visits, to try to be around the house more to support, to assist, to accompany, to be with.

(Continued on page 4)

“Simple Simon Says…”

Goals and Targets

According to the management gurus, goals and targets should be SMART. The letters stand for:

S for Specific

M for Measurable

A for Achievable

R for Relevant or for Realistic

T for Time-based

and goals should usually be all of these things.

“Putting the First Man on the Moon” was a SMART goal. It is very specific and you can “measure” whether it has been done or not. It was just about achievable at the time. It was realistic (man was orbiting the moon already), and it was time-based, in that it was set as a target for the next years of NASA’s space program.

My wife used to work for a multi-national pharmaceutical company. The “SMART” acronym was taught to her as useful in her management role, but I think that it is useful to apply to most other activities.

Setting goals is part of everyone’s life. When you are unwell the goals you aim for will, probably, have to be a lot less challenging and demanding than those you set when you were well. You cannot expect to achieve the same things that you used to be able to. This is the same as when you get older. Start by setting goals that are within your reach. Otherwise you will get down-hearted, and decide not to do anything much with your time.

Don’t worry if you achieve the goals “too” easily. You can always make them more difficult if you are brave, (or foolish), enough. If you have set the goal-posts too wide apart (goal-posts like in Soccer, **not** in Gridiron!), and things are too easy, you can always make things more difficult for yourself by putting them closer together. Also, if things are just a bit too tricky, then you can always make the goal wider. After all, it’s **your** game, and **you** set the rules!

As time goes by, you will, hopefully, feel confident enough to take more things on. By starting at a level at which you **can** succeed, you will be encouraged to try more. The psychologists call this “positive re-enforcement”, which is just doctor-speak for showing yourself, and your family and friends, that you **can** go just that bit further. It’s important to win at this game at first, so don’t set out to change the world! The first targets may be just getting up and dressed without needing help. When I first came home from hospital, I used a walking frame all the time. I slept in a hospital bed in our dining room downstairs (the dining table and chairs had been moved to the garage by friends). Women from the “Homecare” service would come three times a day, and I had to get washed in the kitchen sink (with the women as “escorts” so that I didn’t fall over!). After we had had a stair-lift fitted, and I had shown that I could safely use it when I was on my own at home. I was able to resume a 2-storey life. Two of my early targets were my first bath at home, and a wet-shave (with no ears or noses getting cut off!).

Gradually, you will begin to aim higher, and the people around you will accept you “having a go”. It’s very similar to a growing child beginning to spread its wings. You will, probably, be pleasantly surprised by what you can achieve if you go carefully, and think about things before you try them. You will have to become the “planner” for your activities. For most of our lives, we just get on with things. Not any more. This is a different way of going about things!

Yours,
Simple Simon

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

ECD Through the Eyes of a Mother

By Jeri

Excitement at the birth of a baby girl! Lots of love, nurturing, sharing and caring, I always knew there was something special about Chrissie. Her love of animals, her kindness, her artistic talents and her beautiful mind are all special. Her dad and I wanted to help her develop into an independent, loving and kind adult who could explore all that life had to offer with good health.

All our plans were working out and coming true. Chrissie graduated from college and went on to get her teaching certificate. She married, had a baby girl of her own and remained happy and healthy while touching many lives in a positive way. She made sacrifices along the way to achieve her goals, putting her love for horses and riding on hold. She had a plan!

In her excitement to return to riding and showing her Morgan Horses, she went to see a doctor regarding her hip pain. She wanted to be ready to ride and show again and this was interfering with her goals. She had given up riding because she thought it was aggravating her hip problem. The Orthopedic Doctor was puzzled by what he saw on the MRI and referred her to more doctors and more tests. The diagnosis was a very rare and unusual disease, Erdheim-Chester Disease.

I'm a Registered Nurse and had never heard of this disease. I did research and asked questions to others in the healthcare industry. Many tears. Many questions. Why my child? She was always healthy and careful about what she ate. Why was this happening to my child who exercised and was so kind and giving to others? This isn't fair. More tears. More fears.

She was always healthy and careful about what she ate. Why was this happening to my child who exercised and was so kind and giving to others?

Chrissie is young, just starting her adult life. She has a young family and plans, hopes and dreams. More questions. Will she get to see her dreams come true? Will she see how her hard work has paid off? Will she ever be free of pain? I asked why it could not have happened to me? It isn't fair!

My unconditional love for my daughter doesn't seem to be enough to help my child at this time. I like to fix things but I feel helpless to fix my child. I feel very frustrated at the healthcare system that seems to turn its back on my child because the disease only affects a few. I feel anger as I help Chrissie fight for her health insurance, her right to have access to her medical records so she can take them with her from one doctor to another.

I have great pride in my daughter.

I have unconditional love for her.

I have fears about what lies ahead.

I have anger for a disease with many unknowns.

I am frustrated because I can't fix the disease.

I have hope and faith that I can help my child show her Morgan Horses again and enjoy her daughter doing the same.

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—February 28, 2009

The ECD Global Alliance participated in the global Rare Disease Day on February 28, 2009. As a Rare Disease Day Partner we joined hundreds of other patient organizations, government agencies, medical societies and companies in focusing attention on rare diseases that day. Thanks to everyone who participated!

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.

(Continued from page 1) Erdheim-Chester Downunder

This last is probably the most difficult for me, but most necessary and an element of our lives we’re still growing through: “Charles” is by nature and choice active, doing, going, involved, out and about, participating in this and that and it’s all good work for others but it means I can tend to ‘not be there’ sufficiently. I suppose this is the second message that comes from our experience of Erdheim-Chester, it is difficult to recognise and make the availability-of-self changes that are required for both patient and (in our case) husband.

But there have been marvellous upsides arising through the presence of Erdheim-Chester Disease in our lives too. Tanya says that it is largely because of her experiences that she now finds she can respond with real compassion and empathy to those around her. I find that I love being with Tanya again and that being ‘home’ has truly opened a new vocation for me. We neither wish that things were different or that Erdheim-Chester hadn’t come visiting: this is, as Qoheleth implied, the season that it is time for in our lives.

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!