Me first day on Unit trying to have fun

Me today

A Jedi is Strong with the FORCE!

How it all began. . . .

It was a hot July (for the UK) in 2006. I was happy in my work as an exterior painter climbing ladders most days, which was physically hard work so I put my unusual level of exhaustion down to the heat. However, as the hot days went on my right knee swelled and my feet became painful making high ladder work difficult at best.

A visit to my GP resulted in a blood test, which I was cool with as I thought I'd simply been overdoing it tad trying to make hay whilst the sun shone. Two days later I was sitting in the Haematology dept at my local hospital. The Consultant said I need to do a Bone Marrow Biopsy to confirm my diagnosis but i think you have either Chronic Lymphatic Leukaemia or Myelofibrosis. I whispered to my wife; “I don't fancy a dose of that CLL and he said of the 2 the Myelofibrosis is the harder to treat”.

Two weeks later my MF was confirmed and I joined an exclusive club that no one voluntarily signs up to be a member of.

I was prescribed Hydroxurea later renamed Hydroxycarbamide, which was brilliant and got my blood counts stable but couldn't do much for my other symptoms ie growing spleen, itching, fatigue. But it served me well. The thing is with the drugs they help alleviate some of your symptom burden but they can't stop the underlying MF eating away at you.

A Wonder Drug ?

I trundled along like this until 2014, having been told in 2007 that I had 3-5 years before I would need to consider a more radical approach. But by now my MF was quite advanced being scored Intermediate 1 to 2 on the DIPSS scale. After much ado I was put on the new wonder drug for MF, Ruxolitinib (Jakafi), which I made a promising start, easing my symptom burden and improving my quality of life substantially.

I was hopeful of gaining another few years out of the Ruxolitinib, but alas I wasn't destined to be on the drug for any great length of time as my MF was fairly well advanced and after about 14 months there was no evidence of my spleen reducing in size.

Ruxolitinib can lull you into thinking you're better than you actually are. So what's wrong with that ? You might ask. You look well, you're doing fine, you're managing. Well I could feel small changes in my body, which I guess you become attuned to living with a chronic condition.
I suspected my MF was creeping on undeterred and I could feel it weakening me. I asked my haematologist why no change to spleen but got no helpful response and I asked if I could try a twinning drug approach but this too fell on deaf ears.

Second Opinion

Due to a lack of local expertise I decided there and then to seek a second opinion and was referred to Manchester Royal Infirmary some 60 miles away, which is quite a long trip for me but I guess nothing to you guys with the distances between States.

Following tests and several consultations in June 2015 things ramped up and began moving very quickly. Based on the test results and the fact I'd been on the best drug available I was offered the chance of a Stem Cell Transplant with my Sister as my Donor.

Decision Time

To be honest, the decision for me was a no brainer because I was high risk of my MF transforming to Acute Myeloid Leukaemia, which if occurred would of meant a double chemo' whammy thereby reducing my overall chances of transplant survival. And it was hard even for the expert to predict how much longer I might go before the AML or MF overwhelmed me. So I signed the consent forms, which I found quite hard as I was basically giving permission to be poisoned with chemotherapy and was admitted to the isolation ward to start the process on 27 August 2015.

I was 100% committed to giving it my best shot and my wife was fully supportive.

In At The Deep End

That same afternoon I was whisked off to the x-ray theatre to have my vein saving Hickman Line inserted into my chest. They x-ray to check it is positioned correctly as there's little margin for error. I recall being told to keep very still as my shoulders were moving due to me chuckling to myself. I explained that as they tilted me back on the theatre table I thought I might slide right off into Thunderbird One (for those of age to remember it). Just my way of coping I guess.

Having my Line fitted brought everything suddenly into focus and it became very real. I was in my isolation room, home for the next 4 or 5 weeks but at least I had a TV, which was free to watch on this ward and my wife was able to stay close by for the duration of my stay.

28 August 2015

The day started early with Observations (temp, blood pressure) and a handful of drugs which I would stay on for many months. In the afternoon I started my *reduced intensity chemotherapy*, which lasted about 6 days. This was to clear out my bone cavities ready to accept my Sister's Stem cells. Elsewhere on the Unit my Sister was donating her precious Stem Cells and she excelled herself by donating 6 million, 4 million of which I would need in a few days time. Dunno who counts them ☺ . . .

After the 6th day, all my blood counts were now at zero, hence the need for isolation along with barrier nursing.

4th September 2015 : D-Day

Today I get my Stem Cell infusion. There's no fanfare of trumpets but the nurses in attendance do their best to build up the event. After all its not everyday you get the chance to save your life. The bag of precious Stem Cells which looks just like a bag of raspberry jam is hooked up on my drip stand and attached to one of my line ports. It's just like having a blood transfusion only much quicker and the bag is empty within half an hour. So that's it, all over in the blink of an eye and a tad of an anti climax. I certainly don't feel any different. The new Stem Cells enter my bloodstream and miraculously find their way into my bone cavities and hopefully start working to fire up the miracle of life in me over the next 3 weeks or so.

Dealing with Isolation.

The days come and go and it's easy to think because you're in hospital you should just stay in bed in your night wear. I'm sure some folk do. But I made a point of getting washed and dressed each day in an effort to separate night and day. In the day I'd sit on top of my bed rather than in it because I felt reasonably well overall. And it didn't affect any treatment such as transfusions as my line was easily accessible.

2 weeks in and my counts aren't showing any upward signs , my whites are at 0.2 or so and one night my temperature spikes above 39 degrees. Fortunately, no infection is present and after several hours of cold packs and ice lollies I'm ok. What sticks in my mind is the severe shaking reaction accompanying the temperature as it doesn't seem right to shiver and shake when you're burning up. Also the nurse was trying to insert a needle for a blood test because if she took the blood from my line it might of produced a false reading if the there was any infection in my line rather than in my body somewhere.

Overall the first 3 weeks are manageable especially as my wife is close by in hospital owned accommodation and is allowed to stay with me much of the time.

By week 4 I've had enough and I'm ready to go home. Luckily it's been largely uneventful thus far apart from the high temperature so I'm hopeful of early release. But alas my liver counts are very high due to the effects of the chemotherapy and the drug regime so I'm kept in another week for monitoring. This last week is hard, as I feel ok and am pacing up and down my room to keep active counting steps to 500. It's going well but then in the last few days I suddenly develop Gout in my right foot which lays me up. Much later it emerged that people taking Ruxolitinib prior to transplant are more prone to gout afterwards.

However it didn't prevent my discharge and after 5 weeks I was hobbling home.

There's No Place Like Home

We were looking forward to getting home. My wife had cleaned it thoroughly before we went to hospital because during this initial period you need to take extra care not to get an infection as your counts are very low and you're no longer cooped up in a protective bubble.
However, whilst in hospital one of my Stepsons rang to say he needed to stay for a few weeks. Fair enough but keep the place clean and tidy my wife tells him because Chris needs a clean environment.

Well we landed home by taxi around midnight and upon entering our home my wife isn't happy about the state of the place especially the kitchen. It seems our view of clean and tidy isn't the same as a 25

year old's view. The dishcloth was so *minging* dirty it could of walked. My wife tells me to go up to bed and brings me a cup of tea before setting about disinfecting the kitchen and bathroom areas. It's gone 2AM before she hits the sack but she feels a lot happier.

I wasn't home for long as I needed to attend clinic each week to check progress but it soon became clear my Graft wasn't working as it should be, in fact it wasn't working at all and my counts were at ground level. A top up of Stem Cells was on the cards, as I had some in reserve. But first my Haematologist decided to stop all my Immunosuppressant to see if this action would kick-start my life generating Graft. This stroke of genius worked, and things soon started firing up much to my relief. Happy days.

Graft Versus Host Disease

Until now because the Graft hadn't been working I hadn't had any sign of Graft Versus Host Disease (GVHD), which is usually a good sign that things are happening, as what's left of the old you conflicts with the new invaders. This is often no more than a mild to serious rash (like a burn at worst) somewhere on the body. I had been warned that in a few cases GVHD could be much worse and affect not only your skin but your gut, bowel, liver and in extreme cases can be fatal. But nothing really prepared me for the reality of what happened next.

In late November 15, I became very ill very quickly, and was re-admitted to the isolation unit to be treated for serious Grade 4 of 4 Stages GVHD.

I started with sickness and diarrhoea for 2 weeks solid, my gut and oesophagus were so badly inflamed I couldn't eat. And my vomit was black due to the blood in my stomach, which was scary. Over the next few weeks my skin began to thicken on my hands and feet and was peeling and cracking. I was covered in dry flaky skin and when the nurses changed my bed each morning it was like shaking a snow globe. I lost all my finger and toe nails, in fact on waking I found a toe nail stuck in my backside one morning which made me laugh.

Maintaining weight was an issue and it dropped again and I was in a bad way for a while. But somehow I got through that initial severe attack and by mid January 16, I was just about well enough to go home.

BK Virus

It was fairly short lived as by March 16, I became gravely Ill and couldn't even get off our sofa unaided. I was extremely weak and felt like I didn't have any fight left. My Sister took me back to Manchester and again I was admitted. I had developed a BK Virus which lies dormant until your defences are down then it kicks you harder robbing your muscle mass and appetite so your weight drops away rapidly. You also start passing blood clots each time you pee which is very painful when you're so weak. I was in a desperate way and not expected to survive given I was already battling the GVHD. I was bed ridden and weighed a pitiful 5 stone 10 lb or 80lb.
By some miracle I pulled through and was sent home after 3 weeks. But I was unable to walk, my appetite wasn't good and I was still very weak.

The Long Road to Recovery

I made a slow recovery over the next 2 and a half years and reckon I got back to roughly 70% of the person I used to be. My consultant tried everything at his disposal to help me counter the GVHD from

injections of an experimental type drug, to twice weekly sessions of something called Extra Corporeal Photopheresis, where I was hooked up to a machine which extracts an amount of blood depending on your weight and separates the white cells which are then exposed to Ultra Violet light for a time before returning everything back to you. It was hard to measure how effective this treatment was but I had it for over 12 months and I like to think it played a part in my recovery.

I also needed high dose steroid treatment for the GVHD to save my liver which took a massive hit. The steroids proved to be a necessary evil for me, as they gave me bad cataracts, which required laser surgery and new lens inserted in both my eyes. Also Diabetes for which I needed insulin until I stopped the steroids and my sugars normalised. But worst of all is the Osteoporosis, which led to several compressed vertebrae causing me screaming pain like I've never felt before and costing me 2 inches height loss, and I was only 5 feet 4 inches to start with !!!

My other remaining and troublesome issue is my severe dry eyes caused by the GVHD. I attend a specialist eye clinic every few months and have Autologous eye drops which are made from my blood serum.

I can't write this and overlook the impact my good lady wife made as she was pivotal to my recovery, looking after me 24/7 and making sure I ate what I could, took my drugs etc and helped me wash and use my commode (that's real love for you). Without her unwavering care I doubt I would have made it through. Though it sure tested our relationship at times.

Going Forward

On reading this, you might think what a horror story why would I willingly sign up for that? Well it's a fair question but as you can tell I'm still here to relate my tale. And during the time since my Transplant I've got 3 more Grand-Children come along which is a real blessing and makes it all the more worthwhile.

Also what you need to bear in mind is mine was a rare severe reaction, a friends GVHD consisted only of a small rash on the back of her right hand. There aren't many who experience Grade 4 GVHD, which has a 30% survival. In fact I know of only 2 others with Stage 3/4 GVHD.

Additionally as part of the transplant process the experts are now looking at ways of predicting those at high risk of GVHD and looking at ways of lessening the potential effect before it happens. I was told they learnt much from my experience which was good to know.

The acid test of course is when people ask if I regret my SCT with what happened after. My answer is a resounding NO because without it I wouldn't be here now enjoying life again albeit with limitations. I found out too that I am a mentally strong person.

I now attend clinic once every 6 or 8 weeks and generally feel well most of the time.
To anybody reading this and possibly considering going ahead with a Stem Cell Transplant…

…I would say you would benefit from finding out all you can about it, get as physically fit as you can, even gain a few extra pounds, and having made your decision putting total faith in your transplant team and being 100% committed to the process.

Then go for it.

Regards - Chris