

TUESDAY, NOVEMBER 23, 2004

Here we go...

Well, it seems like we have been waiting for a long time, and we are finally nearly there. Saffy will be admitted into Great Ormond Street Hospital (GOSH) in London on Thursday (25/11/04) to start chemotherapy before having the transplant itself on December 8th.

Thanks to our friends at International Society for Mannosidosis and Related Diseases (ISMRD), Sonja and I (Martin) feel like we know the basics of what we are about to go through with Saffy and how she will feel. The advice from the staff at GOSH has been superb - they really are a world-class hospital. Despite all of this, until we start, we won't really know what the reality of it all will be.

As we find out, we will put thoughts, observations, news and detail on this site. We hope that friends and family will find this useful as we may not have the chance to speak to everyone in person as often as we would like. Please send messages back - we want to be able to tell Saffy who is sending messages. Even better would be pictures, to help her stay in touch with the outside world and all of her friends. I think you can post them on this site, but let me know by message if not and we'll find another way.

We all look forward to staying in touch over the coming months.

Martin, Sonja, Reuben and, of course, Saffy.

posted by Saffron Woolley @ **10:13 AM** -

8 Comments:

- At **12:37 PM**, Anonymous said...

Hi Saffy, Sonja, Martin and Reuben!

We, the Murphys and all your friends at ISMRD, are excited that you have this great opportunity to make life a little more "normal" for Saffy. Thank you for sharing this journey with all of us and the world. The experience you chart will be invaluable to those families who will follow behind, when the time comes for them to decide on the efficacy of a BMT for their child.

Please let us know if there's anything we can do to help, and Taryn and I will be happy to send Saffy some photos from our visit with you in August.

Take care and bon voyage!

Paul Murphy
President ISMRD
The International Advocate for Glycoprotein Storage Diseases
www.ismrd.org

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- At [2:42 PM](#), Anonymous said...

Hugs and prayers to the Woolley family as you embark on this journey with Saffy.

Love

Jennifer, Bill and Matthew (ML3) Simpson
Canada

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- At [8:26 PM](#), Anonymous said...

Dear Wooley's

Our thoughts and prayers will be with you all throughout this experience. We would love to be able to give you all hugs in person but these kind will have to do until we see you all again!!

((((((Martin, Sonja, Reuben and SAFFY))))))

Much Love and endless prayers for Saffy

The Haggett family, John, Brenda & Zach
Syracuse, NY USA

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- At [9:22 AM](#), Anonymous said...

Hello Martin, Sonja, Reuben and Saffy,

I'm so pleased the time has finally come for you. we certainly will be thinking of all of you, and Saffy in particular. Judith and I still remember her as the cutest little English girl we have ever met. Tim and I met Ryan Dernie and his family a couple of weeks ago. He is doing very well about 16 months post BMT. We were taking about you all as we wandered around the Zoo in Auckland, and felt it must be coming close. Our biggest hugs and best wishes. Cheers, John and Judith.

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- At [2:07 PM](#), [Bristol bunch](#) said...

Hello to all The Wooleys - especially Saffy.

Just wanted to say good luck and that we are thinking about you.

Much love

Howard, Henri & Esme

PS. You should put on a link/instruction for people to discover how they register as a BM donor

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- At **4:44 PM**, [AAESumner](#) said...

Good luck to you all.

We are all thinking about you.

Looking forward to another performance by the Keystone Kops next Bonfire Night.

We'll be in touch.

All our love and very best wishes.

Andy, Anne and Ella

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- At **8:53 PM**, [Saffron Woolley](#) said...

Thanks everyone for your messages. We'll pass them on to Saff, who will nod as if she knows what we're talking about, then carry on telling her baby doll to be a good girl and telling us it's not raining outside.

H - good call - anyone who wants to go on the bone marrow register (please do) - start at:
<http://www.anthonynolan.org.uk/ndonors/donor.html>

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- At **5:57 PM**, Anonymous said...

Sonja, Martin, Reuben and Saffy,

Your family has been in our thoughts and prayers. We only wish that we were not an ocean away and could be of more help.

Hugs to all of you!

John, Diane, Katie, Heather and Emily Chika

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THURSDAY, NOVEMBER 25, 2004

D Day (or T -13 Day, to be precise) 25/11/04

So, Saff was finally admitted to Great Ormond Street today. It feels to me like some of the weight has been lifted, even if that sounds a little paradoxical. At least now we are doing something, rather than waiting. Some people less lucky than us have far worse and longer waits for a donor, but I'm not sure Son or I could have done much more of it. We have been obsessing on,

dreading, yearning for and imagining it since February and I don't think we could have done any more. We have jumped now, there's no going back and the parachute *will* open. Of course it will.

For Saff, the coming days will hold an operation that will insert a Hickman line, harvest her bone marrow and take a fibroblast of her skin. The Hickman line is a double tube that will be inserted into her chest. It runs from a major blood vessel, under her skin to a double port that emerges near her neck. The purpose of this is to allow the many (many!) injections and blood samples to be collected easily and to allow chemotherapy drugs to be passed directly into a major vessel, as they cannot be injected into periferal, small vessels (I chickened out of asking why). The bone marrow harvest is a precaution - if the donor bone marrow fails to take (or engraft, as they say), Saffy will be left without any of her own, so the frozen marrow taken from her is put back. That way she has a working immune system while we think about what we do next. The fibroblast is a simple skin sample (taken out with an implement like a corkscrew, alarmingly, but don't worry, it's straightforward and relatively painless) - this is being sent to a leading alpha-mannosidosis specialist, Dr Dag Malm at the University of Tromso*. Can't remember what he needs it for now, but we'll do anything Dag tells us to. I'll look it up if anyone really wants to know.

After that, it's on with the chemo. That's when the fun will really start, by the sound of it, but more about that later.

I took some pictures of Saffy today (sporting her new, short-haired look, in preparation for it all falling out soon, but looking really, really cute, nonetheless) and I should get them back in a day or two and I'll try to put them up here. I'll probably crash the whole internet or something.

All the best

Martin

*Tromso is in northern Norway, of course.

posted by Saffron Woolley @ [9:53 PM](#)

2 Comments:

- At [9:45 AM](#), Anonymous said...

Hi Mart (& Son and Saff),

This is a great idea - among all other benefits pointed out saves you receiving a call from me (& mum) every 5 mins. All the very best for today - of course will be thinking of you.

See/speak to you later

Lots of love and very big hug to Saff

Paula xxxx

P.S. Was really nice to read the message from Paul (Murphy) and everyone else you met via the conference in the US - what a tremendous group of people you met there! (BTW, "Hi Paul - will mail you")

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- At [8:47 PM](#), Anonymous said...

Martin, Son, Reuben & Saffy

What a fab idea - saves you repeating everything a hundred times and it'll help us focus our thoughts. Thank you for including us in your email - we're wishing you all the best.

Nigel, Kerry & Rowan

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FRIDAY, NOVEMBER 26, 2004

T -12. Coming around.

Saffy's operation went entirely according to plan. It was about 2 1/4 hours, with two different teams doing the insertion of the Hickman line and the bone marrow harvest. She is normally groany, moany and pukey after a general anaesthetic, but this time was not sick and, though very groggy and slightly unimpressed, was pretty much ok today. She's a trouper, tough as boots and constantly amazes us.

She now has a double line that literally emerges from the skin of her chest, which is going to take a bit of getting used to for all of us, but it will save countless injections.

GOSH were great as usual - a senior nurse came and took us through care of the line. It actually gets passed down her jugular on the side of her neck into or very nearly into her heart, before the other end is brought out of her chest with two separate tubes that can be used for injections, drips and drawing blood. It will be in after we go home, so we need to know how to keep it clean and germ-free. We have an emergency kit in case it tears or breaks (very unlikely, but if it happens, clamp it twice and get to the nearest hospital pretty quickly!)

As I left to today (Sonja is doing a second night with Saffy - we are trying to limit ourselves to two nights in a row at most - thanks for the tip Mark!) Saffy was actually sat listening to the singing of the entertainer at a party with a plate of crisps and chocolate fingers on her lap. She is in a different ward at the moment (her BMT unit room is still needed by the previous occupant - Saffy will go in before starting her chemo). The party was for a young boy who was at his local hospital and had an unnoticed complication while undergoing treatment there. He was at a normal school and was not particularly unwell. He was clinically dead for some time and now is severely brain-damaged, paraplegic and breathes through a hole in his throat. He has been on the ward for two years, during which time his parents have lived in a house nearby, funded by Friends of GOSH, and have not been able to work. They are going home, but he will need 24 hr nursing care in the family home as he can spontaneously stop breathing.

I'm not trying to scare you or be over-dramatic, but meeting him and his family brought us up short. Great Ormond Street is like that, it really puts your troubles into perspective to meet people who are going through unbelievably hard situations. You think they should just be quivering, weeping wrecks, but they are strong and they gained a kind of wisdom that I hope rubs off a little. Last time I felt like that was in Rockville at the ISMRD conference.

Anyway, sorry to ramble. My mother will take over tomorrow night. Saffy is allowed only three named carers in with her after the chemo has begun in earnest and she has kindly volunteered to

be the other one. It takes a few days of chemo before this happens (it is called being on red precautions) and is triggered when her immune system has begun to disappear.

Whilst we do not want to expose Saff unnecessarily to the chance of infection now (which would put the BMT back) the next few days will also be the last chance for visitors, so family will come in over the next few days. Although everyone is a potential carrier of infections, that has to be balanced with the benefits - for Saffy and those left outside - of seeing each other while there is still a chance.

One last word - I'll talk sometime soon about what we are trying to do to make Reuben's experience as good as possible (we were told that we have two children going through this, and we are trying to heed that lesson). But he's getting the odd treat and tonight's was a trip to see The Incredibles. See this film under any circumstances. It is fantastic. Even if you need to borrow a kid to justify seeing the movie, do it. A definite 5 stars from Reubs & me.

Martin

posted by Saffron Woolley @ **10:01 PM**

3 Comments:

- At **1:00 AM**, Anonymous said...

Hi Saffy, Sonja, Reuben & Martin!

Saffy, you're doing great so far and when you get a chance nod at your dad. He's holding up pretty well so far with this blogging thing for a rank amateur. Not likely to be one much longer, I'd say!

Taryn and I are visiting her grandmother, and my mom, in Virginia. Yesterday, Thursday, we had Thanksgiving dinner out and I told my mother how at that very moment a little girl named Saffron was taking the first step in her new life. We are thankful that your parents were able to give you this chance. Continued best wishes, Saffy, and when I get home tomorrow I'll put together some photos I took when we saw you at the end of the summer.

For now: keep 'em flying!

Paul M in the USA

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- At **9:56 AM**, Anonymous said...

Hi Sonja, Martin, Rueben and Saffy

Our love and best wishes now that you have started your journey. Go for the chippies and choccie bickies Saffy!!

Hugs from all of us to all of you, especially to Saffy from Ryan.

Luv

Sonia, John, Lewis & Ryan Dernie

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- At **2:47 PM**, **Mack** said...

Hi Woolley Family

Just spend the last hour hour reading the blog. I feel very jealous of Reuben Woolley as he got to see The Incredibles. I would love to see that film now but I am stuck in the office, pretending to work. Very hard after three lunchtime pints of beer.

Thanks Martin for the Dylan cd- good choices - we need to have a full debrief.

Love to you all as ever and give that beautiful and brave liitle girl a kiss on her head.

Mack

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SATURDAY, NOVEMBER 27, 2004

T -11 Into our own room

Tonight you have a new, and frankly knackered, scribe. So this is Sonja, fresh from 48 hours (or was it years) of the hospital heat-wave and a diet of chocolate, diet coke and Saffy's leftover chips (much beketchuped).

We moved to Saffy's room on the BMT ward today and what a bloody relief that was. When we got to the hospital on Thursday and found her room on the ward was still occupied, Martin took it like the even-tempered individual he is, Saffy was oblivious, and I sulked, pouted and when all else failed, cried. Today I considered this little interlude a blessing. When I first saw the BMT rooms last month I felt they were modest in size, after 2 nights on a normal ward I thought we'd arrived at a five star hotel (regardless of a broken shower and TV).

I won't recap on Mart's info - just give a brief account of today's news. Saffy started 7 new drugs today in preparation for the start of her chemotherapy tomorrow. I would tell you what they were if I hadn't left my notes at GOSH - it seems that this process isn't going to turn me into an organised Mum. They included gunk to help her liver and chest from the onslaught, antibiotics and an infusion of natural antibodies to help counteract the imminent immune system suppression.

Saffy the Brave seems less affected by the huge things that have happened than the small. She seems unperturbed by large operations but goes beserk if you try and change her dressings. Mirroring this behaviour, I was less upset watching the abuse on her little body (although this is hard) than by her saying "home now please" - which broke my heart.

It was great to see my sister Paula today, who immediately realised that Saffy's bored expression was caused by a lack of physical activity, and the two of them charged up and down the corridors which worked a treat.

Erika (Mart's mum) has taken over the hot seat so I got a chance to see my lovely boy tonight. He

is full of The Incredibles and Ebenezer Scrooge (his role in the school play). Being at home, without Saffy, seems odd and wrong. This will take some getting used to.

Thanks for all the messages - it was really uplifting to know you're watching Saffy's progress.

Sonja

posted by Saffron Woolley @ [9:07 PM](#)

1 Comments:

- At [5:18 PM](#), [Catherine Sumners](#) said...

Hi Sonja,

Love the diary. Thinking about you pretty much all the time. Anything you need (am so close to GOSH) text me and I'll pop over with it. All the Sticky people, my family, Nicki etc are asking after you and send their love. As do Dom, Tilly, Hattie and I. Cxxx

PS Organise smorganise. You're a shoe-in for Mum of the year in my book.

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TUESDAY, NOVEMBER 30, 2004

T -10. The chemo begins.

Well, the phoney war is officially over.

Saffy started Busulphan, the first of her three chemotherapy drugs today. The others will be Alemtuzumab – aka Campath – and Cyclophosphamide. She will be taking these over a strictly-planned timetable over the next 10 days, during which some of the side-effects will start to become apparent. A lot of them will not happen until well after transplant, surprisingly.

Chemotherapy drugs target fast-growing cells and kill them off. This includes blood cells and bone marrow, but also things like hair. This is why BMT patients lose their hair. Other fast-growing cells are found in the mouth, throat and digestive system, causing nausea, vomiting and diarrhoea and she will be given various different drugs to try and control these symptoms, as well as getting pain relief. For the time being, though, she continues to be her old self, and I would be willing to gamble a few pounds that she will still be smiling in 10 days' time. You certainly couldn't tell that she was a chemo patient as she chased Reuben around a table at top speed this afternoon.

The three primary carers (Sonja, my mum Erika and I) sat down for what I can only describe as a tutorial with our assigned nurse today. We had heard a lot of this information before, but everything was really clearly explained and we are actually living it now, so we know what is expected of us, and what to expect, far more now. There is still a part of me that wishes I knew less rather than more, but anyway...

Our routine will include 9-point hand washing with soap and water and alcohol gel. I'm estimating around 25 times a day, based on today. We will save all urine and stools for analysis (I hope

you're not having breakfast right now). Teeth will be brushed 4 times a day, followed by swabbing with mouthwash. A new toothbrush each day. Sterile water only for tooth brushing and drinking. Moisturiser frequently applied to chapped lips.

Sheets to be changed a minimum of daily and placed in the appropriate bin according to the level of soiling. Bath daily. Oil in bath water and all over moisturiser afterwards. Check for any rashes and avoid any water getting into dressings or the Hickman line. There's more (Sonja made 5 A4 sheets of notes), but that'll do for now.

We're moving into isolation (actually green, rather than red precautions as I said before – red is if you have an infection) on T -6 – next Thursday. Things will get really strict then.

I'm off to bed.

Martin

posted by Saffron Woolley @ **3:40 PM**

2 Comments:

- At **3:57 PM**, Anonymous said...

Hi guys,

The day has finally come and we're wishing all the best. I think of our little penguin pilot sitting in his plane with a "thumbs up" sign. Like him, Saffy is "ready for takeoff!"

Keep 'em flying!

Paul, Taryn & Debbie

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- At **5:30 PM**, Anonymous said...

Dear Martin and Sonja,

All of my thoughts and many prayers are with each of you today. Saffy and her parents are so very brave!!! I know this has all got to be so surreal but you will get through it, I know you will.

I'm delighted to hear that Reuben got a little treat for himself and dad too! I will continue to pray that the chemo does it's job in preparing Saffy for the BMT and that the process continues as smoothly as possible.

You two are just as strong as two parents can be so just keep swimming!!!

Lots of Love and hugs!!

The Haggett's, John, Brenda and Zach (MLIII)
Syracuse, New York

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TUESDAY, NOVEMBER 30, 2004

T -9, T -8 Step Change

We've missed a day of posting for the first time. This is down to several factors. Not being able to access the internet from our room was one, but I've sorted that one out now, so we should be a bit quicker replying to emails and messages now. (Best address to mail on is mwoolley13@aol.com)

There were a couple of other reasons though.

The pace of life is furiously fast. I guess the day to day things will settle down into a routine (they already are slowly, to be honest). These are things like the hand washing, bed changing, oral care, moisturising, hand washing, sterilising, bottle preparation, hand washing, skin care and stock taking of in-room essentials. You have to remember to wash hands a lot, too. In addition, you (as well as everyone else) have to put a plastic apron on when entering the room and remove it when leaving. One of the drugs has also upset Saffy's stomach, making her have very frequent and rather difficult bowel movements. Each time this happens, you have to put disposable gloves on (this is to protect you from chemo drugs as they are passed out of the gut and bladder) and the use of bed pans, saving of nappies, hand washing etc all takes a certain amount of time. At one point yesterday morning, that was all we did for about an hour - by the time I finished, she started again. On one particularly fun occasion, she started again before I'd finished. She had been in knickers until then, but seemed actually to resign herself to the fact that that wasn't working and graciously agreed to compromise on pull-ups after she'd seen me throw about 10 pairs of M&S briefs away. I think I developed a new level of respect for my daughter at that moment.

Anyway, enough of toilet matters. We started another chemo drug today - Campath. This is quite a nasty one, infused into the blood through the Hickman line over about 8 hours from a drip. Possible side-effects are high temperature, rashes, nausea and convulsions, so she gets a premed of paracetamol, piriton, pethidine* amongst other things. This controlled all of these symptoms effectively, as well as knocking Saff out for part of the day. She was fantastic about staying in bed most of the day and having tens of syringes-full of drugs through her line and orally. She is still on Busulphan until tomorrow as well, so was nil by mouth for 2 1/2 hrs for each dose, so she had a lot to put up with today. The consultant says she absorbs Busulphan at a perfect 'moderate' rate, too. That's my girl.

Sonja is staying in tonight - the rota seems to be working out well and giving us time with Saff and Reuben without them missing us too much or getting too jealous so far. Early days, but fingers crossed. Touchingly, they seem to be genuinely missing each other. Reuben wanted to know how they got her 'wiggly lines' out of her chest. I explained it while he was in the bath tonight and he thought it was brilliantly gross - I expect all the boys in class 1A will have heard about it by lunchtime tomorrow. (He doesn't talk to girls. Except Lizzie, but that's a different story...)

We also met a play specialist, who'll buy new toys for Saff based on what I told her she likes playing, a dietician, who'll stop by tomorrow to discuss favourite foods for her when she moves onto green precautions on Thursday and gets all of her meals specially prepared - she will try and give Saff a diet of favourites to keep her eating when she starts to feel worse. We even had a clown, who, even though she couldn't come into the room, did magic tricks and funnies for Saff through her window onto the corridor, which went down a treat. She's also had grandparents,

aunts and uncles in recently, so she's feeling quite at home just now. It may change when she's on green precautions and no one is allowed in, but we'll cross that bridge...

Sorry - bit of an essay this time. Two days' worth!

Thanks so much for the messages, though. Now we can pick them up in the room when we're together, it's even nicer to hear from you all, so do keep posting messages, gossip, jokes whatever and let anyone we haven't told have the address.

Cheers

Martin

*It's the one they can give at childbirth - that's why it rings a bell

posted by Saffron Woolley @ **9:00 PM**

1 Comments:

- At **3:56 AM**, Anonymous said...

Martin,

Well, I've learned one great lesson so far: wash your hands before, during, after and in-between meals. Then wash them again! I think I've got it now!

The blog seems to be doing very nicely, in spite of the need to stay focused on more important things (don't forget to wash those hands...). I know there will be stretches when, despite what routine you think you might eventually settle in, it's just too much to keep up with on a consistent basis. Don't worry about your poor readers, though, just keep little Saffy happy and cared for (no doubt at all that that wouldn't be the case).

So, gossip and jokes is it you want? How about this one:

Q: Why was 6 afraid of 7?

A: Because 789!

On a serious note: today I bought a house and we will be moving in about two weeks. Taryn and Debbie are both thrilled, as am I. I'll email you the details later.

Hi Sonja, Hi Saffy, Hey Reuben

Cheerio...

Paul

-

WEDNESDAY, DECEMBER 01, 2004

T -7 Raspberry Nipple

Tonight's entry will be both poo-free and brief. The former because I think Mart's put you in the picture and I've had quite enough of it for today. The latter because I'm v. tired (again), I've only got 20 pages of Persuasion left to read and it's winking at me from my bedside table. Now I know this sounds selfish, but come on, it is Jane Austen.

Saffy continued to be remarkably well today (surprising to me, not necessarily the nurses) but appeared to be suffering from an acute attack of boredom. This would have been easily deduced by anyone entering her room and discovering that, scornful of my attempts to excite or even distract her, we'd resorted to a few lengthy rounds of who can blow the loudest raspberry. It was close.

We were given useful guidance from yesterday's nurse about when Saffy is likely to start feeling worse. Most children deteriorate shortly after the chemotherapy has finished and therefore shortly after transplant (T+3ish – or around Dec. 11th in your real world). They don't improve until the donor marrow starts to re-boot their immune system (usually 2-3 weeks later). The dangers then change to infection or rejection – well we'll be needing something to worry about.

Medically speaking nothing new was introduced today. Chemo 1 (Busulphan) is about to release it's final dose and Chemo 2, (Campath) is at the half-way stage of a four day mission. Chemo 3 (Cyclophosphamide) has to wait until Friday for launch.

I was a convincing winner of fool of the day here on Fox ward. I drew our nurse's attention to a worrying red mark just under Saffy's Hickman Line entry wound, then realised when I took a step back that it was her nipple. Still I'm sure the nurse kept that that one to herself.

Must go and check that Anne Elliot still pulls Captain Wentworth.

Thanks for the many messages, we really enjoy reading them.

Sonja

posted by Saffron Woolley @ [10:36 PM](#)

4 Comments:

- At [2:32 PM](#), Anonymous said...

Hi Martin, Sonja, Reuben and of course saffy

its vicki here your hard working inserts secretary at News int (haha!).

sorry i havn't posted sooner its real hard to think what to say at times like this so i'll start by saying i hope your all as well as can be, just been reading the full blog and i have to say i feel dreadful for complaining about something as trivial as a headache yesterday!!

things here are the same as usual really not much changes hey.

cant think of any jokes apart from a chris wyatt classic that always springs to mind

Q: what did the Number 0 say to the number 8??

A: Nice belt!

silly i know but if you knew mr wyatt it may make you chuckle?!?!

i wish you all the best and hope everything keeps going to plan with saffy's treatment, i think i speak for everyone when i say out thoughts are with you and we hope to hear lots of positives soon.

stay strong and will keep following saffy's progress (this blog is a great idea)

well take care of each other, i suppose i better get working again?!?!

much love
vicki x

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- At [4:35 PM](#), Anonymous said...

Hi Sonja and Martin

Its Jacqui (cousin) here - I've only just got access to this website - what a great idea.

To date, Mum has been keeping me updated about whats going on, but I just wanted to let you know that I'm thinking of you all and I hope Saffy is doing OK

Also, if you ever need my services to look after Reuben or run any errands for you, please don't hesitate to ask as an ex-Palmers Green resident, I don't mind travelling to North London!

Take care, J xx

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- At [5:45 PM](#), Anonymous said...

Hi Woolleys. Mack here. You would think after 10 years in IT I would be able to post a comment fairly easily. Think again. I was only thinking yesterday that I should read my first Jane Austen novel since A level English. Not sure I even read that to be honest. Just copied other people's notes and watched the film.

I am not sure if you ever got my first message but suffice to say, love to you all and if any of you (probably not Reuben or Saffy) can sneak out for a beer while at the hospital, I would love to buy you one. If you want to pay, even better. Mack

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- At [10:09 AM](#), Anonymous said...

Hello Woolleys, just to say hi and that we are thinking of all of you. Also to reiterate Jac's offer - we are always available for babysitting, trips to the cinema, McDonald's etc with Rueben. I am working on Holloway Road anyway, so is just up the road for me.

Keeping fingers crossed for Saffy, and keep up the blog - it's great!

Lots of love Helen xxx

FRIDAY, DECEMBER 03, 2004

T-6 Slightly choppy waters

I feel that I tempted fate with yesterday's entry. A few hours after posting how well everything seemed to be going, Saffy woke with a really high temperature. The nurses don't give paracetamol immediately as they don't want to mask an infection, but following two high readings they gave her a dose and asked the doctor to look at her.

Saffy had shown signs of a mild rash following her second dose of Campath and this now spread all over her body. The nurses described it as quite mild (I would have opted for "livid"). The doctor believed the rash was a reaction to the chemotherapy, but the temperature, which didn't start to rise until over 10 hours after the Campath infusion had finished, probably wasn't. This meant Saffy could have an infection and blood was duly taken and sent to the lab. It takes up to 48 hours to see if a bug grows, but the doctor decided not to take any chances and placed her on antibiotics straight away. They also gave her more Piriton (antihistamine) to reduce the reaction to the chemotherapy. The medical team described this as covering all the bases – i.e. regardless of what is causing the fever they are counteracting it.

Saffy responded extremely quickly following the paracetamol and was bouncing around her room in no time. At about 11am she had to be hooked up to the Campath again and became increasingly subdued over the course of the infusion (now about four hours). By the end Saffy was lying shivering on the bed with a temperature over 40 degrees. I felt very anxious, but our wonderful nurse remained extremely calm and rated her own concern as "mild". Sure enough another dose of paracetamol did the trick and by the time I left the hospital at about 6 last night she was back to giggling and playing boo from under the bed sheets.

Mum and Dad, Paula, Martin and I were all in the hospital for most of yesterday. As Saffy was suffering under the strain of the chemo, we were busy scrubbing all our possessions with alcohol wipes for the transfer to her isolation room. Only Martin and I could enter it. The rules are now much stricter. Only Saffy's three main carers are allowed in the room and Saffy can't leave it. Aprons are now compulsory and must be changed each time you enter or exit the room. Hand-washing is now almost continuous!

I'm writing this from the comfort of home on Friday morning. I have to keep resisting the urge to get up and wash my hands! Home time means laundry time, and our overworked washing machine and dryer are providing a noisy accompaniment to my musings. I'm not going to hospital today, this will be the first time I haven't been in and it's very odd. Martin keeps reminding me that

this is a marathon not a sprint, and as the first week has exhausted us we're now considering the best ways to conserve energy. Not going to hospital on a day off is hard but wise.

So I'm putting up the Christmas tree today as a surprise for Reuben when he comes home, then cooking a nice dinner for myself and Martin, who will be home later when Erika's shift starts.

We know lots of you are struggling to leave a message on the site, if this happens, please just contact us via Martin's email, mwoolley13@aol.com.

Wow this is long.

Sonja

posted by Saffron Woolley @ [11:23 AM](#)

1 Comments:

- At [11:32 AM](#), Anonymous said...

Hi Sonja,

Glad to read that Saffy's temperature came down. I hope she continues as well. It seems funny that we can't see her now for at least a month. You'll be glad to know that the prayer campaign is continuing!!

Love to all of you and a very special hug for Saffy.

Love M.

SUNDAY, DECEMBER 05, 2004

T -5, T -4 What's your poison?

Since I've last been on, my mum has been on duty. This coincided with the start of the third and, I suspect, least pleasant of Saff's chemo drugs, Cyclophosphamide. (They all have such unpleasant sounding names, even.) Cyclophosphamide is the one most likely to cause sickness and also is an irritant to the bladder, causing blood in the urine. Consequently, Saff has to go to the loo every 2 hours, night and day and was also sick for 2 hours last night. This combo meant that I don't think my mum actually got any sleep in the last 48 hours, so she'll probably sleep from now until the next shift.

Saff's been on a drip (or 2 or 3) for the last 72 hours and has at least another 24 to go. This means not leaving her bed during any of this time. Her carer cannot leave her at unattended either, so we are trying to make sure that there are 2 people here during the day whenever we can as there are lots of reasons to leave the room. We have to get all of our supplies from bed sheets to sterile water (used for everything from drinking to botty wiping) to clean bed pans. From time to time we need to eat, too! (Especially me). Nurses are happy to sit in, but some times are busier than others.

Nausea is being controlled by 2 main drugs. Ondansatron acts on the nausea centre of the brain and Maxillon acts by encouraging the muscle movement of the gut to keep food moving through. If these are not enough, another will be added. If that doesn't work, they'll 'start from the beginning and find a combination that does'. Controlling vomiting is a priority as it, along with the diarrhoea, it will dehydrate Saff and stop her getting the chemo out of her system. (As well as not being much fun, obviously).

Saffy's hydration levels are also monitored really closely. Everything she eats or drinks, wees or poos, is recorded to see if she is negatively or positively hydrated for the day. Depending on the result, she'll get a saline drip or a diuretic to restore the balance.

If you're thinking this is all pretty bad, don't get too concerned. She is also Dr Saff, who with her medical kit, examines and treats everyone from Baby Lucy (thanks for the perfect gift Tony & Anne!) to her own doctor. She's full of it, just like she always is. Added to which, on one side of us we have a South African couple who have had to move themselves, 2 other kids and grandma over to England for 3 months (and counting) so their son can get treatment unavailable out there. They are managing to work long-distance and school their kids at the same time. Just down the hall is the guy you might remember from the papers recently who needed – and found – bone marrow donors for all 4 of his boys for a genetic condition of the immune system. He's in with son number 2.

All of which reinforces that having as much time as I need from work is an enormous blessing. I'm sorry for anyone having to do more work as a result - I'm really grateful. There are also 2 kids - the one in hospital and the one at home - getting the best chance to overcome a lot of bad luck and live near-normal lives as a result. Thanks a lot from all of us.

See you soon for another thrilling installment of our hospital drama.....

Martin

posted by Saffron Woolley @ **4:09 PM**

3 Comments:

- At **1:34 PM**, Anonymous said...

Dear Dr Saff

I hear you've now joined the medical profession so thought you ought to brush up on your Dr, Dr Jokes:

Man: Doctor, me leg keeps talkin' to me.

Doc: Don't be ridiculous!

Leg: Lend us a fiver!

Man: Told you.

Leg: Give us a tenner!

Doc: My god!

Leg: Eh Doc, can you spare 20 quid?

Doc: I know your problem. Your leg's broke!

Best wishes

Kerry

xxx

-

- At **9:04 PM**, **Saffron Woolley** said...

i'm sorry Kerry, but that's a stinker. Almost as bad as the man going to the doctor with a steering wheel on his willy and says 'take a look at this Doc, it's driving me nuts!'

-

- At **2:15 PM**, **sumant** said...

Hi Safi, Martin and Sonja

quick joke...

man goes into a vet and says his parrot's not well..

vet takes one look at it, stiff as a board and says it's dead.

Man disagrees, and wants second opinion...

Vet tuts, goes into next room, and brings back a cat.. The cat paws the parrot, looks up at the vet and shakes its head..

"You see, it's dead," says the vet..

The man still isn't convinced..The vet tuts and goes next door, and brings back a labrador..

The dog paws the parrot, looks up at the vet and shakes its head..

"You see, it's definitely dead," says the vet..

"OK", says the man "How much do I owe you?"

"2000 pounds" says the vet.

"2000 popunds??!!!" says the man.."It's an outrage!"

"Well it was ten pounds", says the vet, "But then you asked for the cat scan and the lab report..."

-

MONDAY, DECEMBER 06, 2004

T -3. How did we get here?

Saffy is currently sleeping soundly after a good day. Having had her sleep interrupted by two-hourly nappy changes and by sickness last night, she was pretty tired and was actually asking to go to sleep by about 7pm. If you know her well, you'll understand how unusual this is!

So I thought, as I'm now getting addicted to this diary-therapy and as there have been several people who have actually said they are reading this, that I'd just take a minute to cover why we are here and what happens next. Some people seeing this know plenty more about this subject than I do – please feel free to correct me. If anyone wants to learn more, visit

www.mannosidosis.org

Saffy has a disease called alpha mannosidosis. It is not very frequent. To put it mildly. You hear phrases bandied around such as 'ultra-orphan' to describe the rarity of conditions like this. I read in a booklet for a rare cancer that affects around 1 in 6000 kids that a GP is likely to encounter only one case in his working life. Alpha mannosidosis is reckoned to affect about 1 in 1,000,000 kids, although no one really knows for sure. It is one of a group of conditions called storage diseases. Storage diseases are genetic in origin. Both parents must be carriers and even then there is a 1 in 4 chance that any child will have the disease.

The term storage disease relates to the fact that, because of a missing enzyme, the body cannot carry out a basic function that occurs within all of its cells. Part of the cell (the lysosome) acts as a kind of mini stomach, breaking down the basic building blocks of food and the products of your metabolism into parts small enough to become the basic building blocks of the body. This function is carried out in pretty much every cell, wherever it is. In alpha mannosidosis, an enzyme called, appropriately enough, alpha mannosidase, is not produced. The basic sugar (very basic form – don't think Tate & Lyle) that it works to break down (along with many other enzymes) is an oligosaccharide (can you believe that word got through the spell check? Impressive). Because the oligosaccharide is not broken down, it's not small enough to leave the cell and has to stay – or be stored – there. Hence storage disease.

This happens all over the body, from brain to liver to bones to skin and basically everywhere else you can think of, and slowly, over time disrupts the function of that organ. Sufferers are affected on a broad spectrum of severity – Saff is towards the least affected end. Whilst all diagnosed kids with alpha mannosidosis will get treatment for all of their diverse problems, the disease has no cure. The only effective therapy is the bone marrow transplant. Saff is lucky enough to be relatively unaffected at the moment – partly because she is a mild case and partly because the man at the top of our Xmas card list, Professor Surtees at GOSH, made an early diagnosis (there were a lot of other important people in getting us to him in the first place and since).

What the BMT will do (if the procedure goes according to plan) is replace Saff's deficient marrow with donor marrow that can produce alpha mannosidase. This will stop further storage and will even start to clear some of what has already accumulated. The effects are quite quick to show in the body. In the brain, the new enzyme has to work its way across the 'blood-brain barrier' – a defensive mechanism there for the brain's own protection and this can take up to 2 years, but it will get there eventually.

So, (almost there now) what will this mean for little Saff?

The main points should be (and no one offers any guarantees on this):

Brain – by 2 years post-BMT, any deterioration will be arrested (fingers crossed there's not much between now and then)

Hearing – not effective, she'll always need hearing aids. The plus side is her aids are fantastic and she gets on with them really well

Enlarged liver – should actually shrink back to normal size

Spine – the current curvature remains but will be treatable as a normal orthopaedic issue

Muscles, heart, eyes, throat, immune system – touch wood, none of these should deteriorate

Bones and joints. These will probably deteriorate a bit faster than in an unaffected person, but nothing like as fast as they would without the BMT

Sonja will no doubt tell me I've made a glaring omission, but I hope you understand now why we felt we had to take this risk for Saff. Hopefully she'll understand herself one day.

I hope that makes a bit of sense.

More from Planet Saffron in a day or two.

Martin

posted by Saffron Woolley @ [10:19 AM](#)

1 Comments:

- At [2:14 PM](#), Anonymous said...

Saffy

Hope you had a better night's sleep than last night? But one small point can you please keep your snoring down it's upsetting the other patients and nurses, have more consideration for your fellow man, anyone would think you were ill or something.

Your Dad tells me you're being very brave (as we expected) and that you're getting lovelier & prettier by the day, again no surprise there, you put the rest of the norf London ladeez to shame. Chloe, Jared & myself are all thinking of you, and we want to send you maximum love, hugs & kisses through the digital ether, which your Mum & Dad can transfer into real ones in our absence. We can't wait to see you in the New Year and hope you'll visit us at the sea-side when the spring and summer finally arrives. We'll obviously expect you & Reub to babysit whilst the adults disappear to the pub, are you cool with that?

Following in the vein of previous comments here's a very old but funny joke (from a fezd up T. Cooper);A man phones the doctor's surgery & says

"I'd like to see a doctor?"

The receptionist answers, "Which doctor?"

The man replies "No, I want to see a real one"

Boom Boom, Ho Ho etc

Toodle pips for now Saffy, all our love

Paul, Chloe & Jared XXXXXXXXXXXXXXXXXXXXXXXXXXXXXXXXXXXXXXX

-
, DECEMBER 07, 2004

T -2, T -1. The end of the beginning

Well, Saffy had her last dose of cyclophosphamide yesterday.

Although we are not out of the woods (just about to enter them!), I can't say I'll miss the sight of a nurse coming in with a large vial wrapped in black plastic - cyclophosphamide is light sensitive - with yellow 'warning toxic' stickers. They also wear gloves, arm guards and a visor. All of which could all freak you out a little, if it wasn't for the fact that they really know what they are doing. Still, they won't be infusing poison into our girl anymore, so we'll take that as a positive. If this year has taught us something, it's probably to find positives where you can and enjoy the little victories; so this counts as one.

Some of the drugs she is having now are to control symptoms such as nausea, pain etc. We are expecting quite a few things to be happening about now. Saff will be getting symptoms of mucositis - inflammation of the lining of the digestive system. In the mouth and gums this can be alleviated to an extent by the oral hygiene routine, but mucositis affects the lining of the digestive system from mouth to the other end and beyond the mouth this is more difficult to treat with mouthwash! We suspect that her throat has been sore and she is now starting to refuse food and drink (as well as tooth brushing and mouthwash swabbing).

The next step to help get oral medication and liquid into her will be a naso-gastric tube (into her stomach through her her nose). Commonly known as an 'NG' around here. Along with the Hickman line into her heart, this will look unpleasant, but will make giving liquids and oral medicines so much more straightforward that it is far more a positive than a negative. The only problem is getting it in as kids can often get upset by this. We are trying to make sure we have spoken to Saff about most of the obvious things that will happen, so they are not shocks (at the moment, she thinks losing her hair will be a big game). We're trying the approach of letting her know about the NG shortly before it happens so it's not a shock, but she doesn't dwell on it too much. Time will tell if this is the right approach.

As for other drugs, (they'll be more detail on these a little later) another new addition is to add Cyclosporin. This will help Saffy prepare to fight Graft vs Host Disease (GvHD) which, along with infections, is our new main enemy. There is no chance at all that Saff will not get some of both of these, but we're hoping for skirmishes rather than pitched battles. GvHD basically means rejection (as with an organ transplant), but there will be plenty more about this as we move into T+ days.

Tomorrow (Wednesday) is transplant day. But it's not really a transplant. Tune in to the Saff Channel soon to find out why.

Don't worry though. Everything so far is according to plan. Our nurses have not got to more than 'mildly concerned' at anything yet and they have done this lots of times. Saff, of course, is still the gorgeous game-playing, grinning minx that she always has been, just a little bit less bouncy.

Martin

PS. Thank you so much for the messages/emails/cards & gifts - Saff & the rest of us really get a lot out of them and it makes all the difference.

PPS. The quality of the jokes is questionable, but I know there are bound to be some crackers about to come in from all those Xmas nights out...


posted by Saffron Woolley @ **9:15 PM** -

3 Comments:


- At **6:18 PM**, Anonymous said...

Mack here. I am addicted to the Saff channel. The pics are a great addition to Queen Saff's site. It's a real page turner and you both write so well. The Woolleys must all be flushed with success after Rodney's victory on I'm a celebrity get me out of here. Let's be clear: Rodney is Joe Pasquale, not just looks and sounds like. Is. Quite literally. I want to send news or a joke (not both) but other than the fact that I am now sharing the house with a rat once more - and yes, quite literally, I have no news.




Saff with her new haircut 
THURSDAY, DECEMBER 09, 2004



More Saff 

THURSDAY, DECEMBER 09, 2004



Yet more Saff 

THURSDAY, DECEMBER 09, 2004

T +0, T +1. The most important little pink bag of Saff's life

Saff has a penchant for bags and for pink. But today she got the the little pink bag that we hope will last when all of her others are in the bargain basket at the charity shop.

If you are anything like we were, you won't quite understand what a bone marrow transplant actually is. A few short months ago we had to admit our lack of understanding to Saff's consultant and ask for a basic explanation. I had pictures running through my head of open bone surgery, injections into bones all over her body and even more macabre delusions.

So, here is a potted guide to what a BMT actually is. This should come with a health warning as we still aren't exactly experts.

First of all, a bone marrow transplant is not really a transplant. It is a transfusion. It looks like a bag of pinkish blood that she gets through her Hickman line in under 1 hour. Basically, marrow (the mushy, magic bit inside your bones that contains stem cells where - amongst other things - blood cells are manufactured) is syringed out of the donor's pelvis. The donor has to be matched to be compatible with the recipient - the match is for an HLA (don't ask - we don't know) tissue type; the donor and recipient can have different blood groups*. The best type of match is from a sibling. Unfortunately, Reuben and Saffy are incompatible.

From an unrelated donor, initial match is sought first on an initial 6 point scale, which is then extended to a 10 point scale and finally a 12 point scale if you have a choice of 10/10 matches. Saffy's donor is 11/12, which is a pretty good match.

The chemotherapy that Saff has had is to kill off all of her own bone marrow, which cannot produce the required alpha-mannosidase enzyme, to make room for donor marrow which can. (Radiotherapy, or high-dose x-rays, is an alternative way of killing bone marrow). In cancers, killing bone marrow is a side effect of using chemo to kill the cancer; with genetic conditions like Saffy's, you want to replace her marrow with a better version.

The plan is for the new marrow to occupy the gaps where Saff's used to be. This is known as engraftment and that will be one of the key words from now on. The problem is that bone marrow is, by definition, full of white blood cells, the primary purpose of which is to fight foreign matter. All of Saffy is foreign according to the donor marrow, so we should expect a fight. This is the Graft vs Host Disease (GvHD) that we mentioned before. There is little chance that it won't happen (particularly as we don't have a 12/12 match or a related donor), but the medical team are expecting it and have won plenty of these battles before. Winning the battle means controlling the symptoms of GvHD and achieving engraftment.

We are actually writing this on day T +1, my mum is on duty, and this is the day the bad symptoms are supposed to kick in. Saff should be feeling very ill, have sore mouth, throat and stomach, and be quite nauseous. My mum reports she has eaten chicken nuggets, toad-in-the-hole, cake, an orange and toast today and taken all her oral medication. The NG tube has been put on hold for now. Her condition will get worse, but she continues to be remarkably robust for now. Go Saff.

As expected, with depleted marrow, her red blood cell count had fallen and she had a blood transfusion. This put a lot of colour back in her cheeks and may be a reason why she is feeling better and has her appetite back.

Although we talk about her being in for a bone marrow transplant, she is actually in for enzyme replacement - BMT is the means to this end - and that can't happen until the new marrow is engrafted successfully. In other words, T +0 is a bit of a non-event in itself and there is not a cause for celebration yet. The (pink) champagne will come out when her blood counts show this has happened and we've beaten infection and GvHD.

Well done for reading this far - we were trying to be brief, honestly!

Take care

Martin & Sonja

* Quirky fact - Some time after all of this is over, Saffy's blood type will change to that of her bone marrow donor. Weird.

posted by Saffron Woolley @ [10:25 PM](#)

3 Comments:

- At [4:07 AM](#), Anonymous said...

I have to ask, what is a toad-in-the-hole :)

Hugs to all of you!
The Chika Family

-

- At [9:21 AM](#), Anonymous said...

Hello Saffy,
and Martin, Sonja and Reuben,
and all the others out there following this saga.

I've followed this with great interest and read ALL the postings and comments. Now it is a calm and mild Sunday evening in Petone (abbreviated and Anglised from Pito-one, the original Maori name for this area), across the harbour from Wellington, and the site of the first organised immigrants' arrival in New Zealand, on 22 January 1840.

No, this is not going to be a history lesson - not that sort anyway. I thought I'd add a few comments to the very good explanations Sonja and Martin have give about Bone Marrow Transplantation for Alpha-Mannosidosis, and try and make this posting informative for those whose lives have not been immersed in Glycoprotein Storage diseases in recent years, and perhaps a little enlightening even for some who have the experience.

Some time in the 1950s or 60s artificial insemination became the buzz in cattle breeding and a prize bull from England was used to inseminate huge numbers of Aberdeen Angus cows here in New Zealand. Unfortunately this bull turned out to be a carrier for Alpha-Mannosidosis and before you knew it, nearly 10% of this breed were carriers of this storage disorder. There were major economic and practical implications. Try handling cattle and you know you have a job on your hands at the best of times. Try handling one with the temperament of "A-Man" and you have a very difficult task, let alone the other complications of the disorder.

Fortunately back then, we had a very bright young Veterinary Pathologist at Massey University, Bob Jolly, who identified the disorder, worked out an enzyme assay test for carrier status, and set up a carrier detection programme for the herds based on the carrier detection programme for Tay-Sachs disease (another Lysosomal disease) common among the Ashkenazim Jewish population.

Bob was very successful with this work and effectively solved this problem in the cattle herds by enabling culling of the sires. (Don't worry Martin, the rules for us humans are different). But that was not the end of the story by any means.

In one case they discovered a carrier cow was pregnant with twins - one affected by A-Man and one not affected. Twins are very rare in cattle, and the characteristics of this pair were very informative. Cell transfer across the placenta meant the unaffected calf gave some of its cells to the affected calf. Nature's own bone marrow transplant!!! (or perhaps stem cell transplant would be more accurate but we won't quibble over the details). Bob wrote up the results and published them and an eager young scientist from New York, Steve Walkley, took an interest. He visited Bob and discussed the work and then tried a BMT in some cats that also were naturally affected by A-Man. The very important aspect of his findings was demonstrating enzyme activity in the brain of the transplanted cats. This gave scientific evidence for the theory that a BMT might correct physical and neurological symptoms of the disorder, or at least minimise some of the symptoms and improve health and quality of life for the affected person. This was not the first BMT for one of these storage diseases - an Australian scientist had demonstrated benefit for a dog affected by Fucosidosis, another of our Glycoprotein storage disease group. But it was a very important step for all the disorders and A-Man in particular. BMT was on the agenda.

The rest is history, as they say. More than 20 BMTs (or stem cell transplants from Cord blood - similar thing) have now been carried out on children affected by A-Man around the world, and the success rate has been very high. My guess is that Saffy's BMT would be around number 21 or 22 of cases transplanted for this disorder.

It is sometimes a sad thing for Judith and I to realise that this treatment was devised too late to benefit our twins, Timothy and Hollie, both of whom are quite significantly affected by A-Man, and who celebrated their 30th birthday last month. It was also sadly fascinating in 1999, the year of our first Lysosomal conference and the year we learnt about and met Bob Jolly, to discover all this information about his work and to realise that tens of thousands of A-Man carrier tests for cattle were carried out just ten minutes drive from our home, yet we didn't even have the correct diagnosis till they were age 15.

But our mixed feelings about that history lesson are overwhelmed by the very good feeling we get knowing that our little country at the end of the world, and one of our top scientists, laid the groundwork for the treatment that offers such hope to Saffy, the cutest little London girl I have ever met.

Now let me indulge in a significant moment. In September 2003 Lysosomal Diseases New Zealand had its very first conference in Auckland and we invited retired Professor Emititus Bob Jolly to give a talk on animal models and their importance for knowledge of the human condition. At the very end of the conference I was talking to Bob in the Hotel foyer and was able to point out to him the little boy running around with a big surgical mask over his face. I told Bob how much we valued his work and how important it was to families, and then told him that young Ryan Dernie was about case number 20, just recovering from his transplant at Starship hospital. Bob likes to think of himself as a tough sort, but he could not disguise the tear in his eye. Martin and Sonja, you can add that to the experience of meeting Steve Walkley at the Washington conference early this year and you should now feel very well connected to the key scientific brains that led to Saffy's treatment.

End of story? Not quite. Bob went on to find several other natural animal models of Lysosomal disease in New Zealand's farm animal population, and there are sheep with Batten disease and a Huntaway dog with Sanfilippo disease, currently adding important information to the collective knowledge. He has also identified sheep with Popme disease way up in the Lindus Pass in the South Island high country.

These are some of the reasons we (NZ Organisation for Rare disorders) are setting up a biobank here in NZ, with a priority to collect and preserve these important natural animal models, and with the aim of organised efforts to find more of them. Who knows what great opportunities there may be for others like sweet young Saffy in the future, as a result of the work of dedicated researchers like Bob. We can't change the past, but we can put effort into making sure there are more opportunities for kids like Saffy, to get treatment for these insidious diseases in the future.

If any of your non-lysosomal friends and acquaintances have hung on to the end of this, Maritn and Sonja, I

wish them well and hope they will hug their friends, partners, and kids and affirm that even when life seems bad, it is good. If they have fallen asleep at the PC after my rambling story, then all I can do is wish the two of you, and Reuben and Saffy, all the very best for the coming weeks. You are all in our hearts.

Regards,
John and Judith Forman
and Tim and Hollie
john.forman@xtra.co.nz

-

- At **4:14 PM**, Anonymous said...

Just to say that your description of the BMT was really useful to me Mart, and the posting by John and Judith was fantastic - very informative (and also very moving -I'm so incredibly sorry for parents with older A-Man children for whom BMTs were not available, but also very moved by their generous unfailing support).

Thanks John and Judith.

From Paula (Sonja's sister)

-

FRIDAY, DECEMBER 10, 2004

T+2 A word from Reuben

hello i am saffy's big brother reuben i feel very sorry for my sister i have been making a lot of things for her.

i am school council! so it means that I go to special meetings and I tell either the headteacher or the deputy headteacher what would make the school better.

I was also the main character in my Christmas play, A Christmas Carol. The main character is Scrooge. He's a mean and miserable man - who hates everyone and is nasty to them and hates Christmas.

My lessons are very good in school. On the 10th December at school I made a moving santa using split pins. I also coloured in Christmas pictures a lot at school.

Goodbye

And I wish you a merry Christmas!

Love from

Reuben

posted by Saffron Woolley @ **7:15 PM** -

2 Comments:

- At [9:29 AM](#), Anonymous said...

Hello Reuben,

A special note for you too. I'm sorry we did not get a chance to meet you when we were in London, but we heard such good things about you. You will have to be strong through this time too, as you will not be able to see your sister for a long time, and mum and dad will be very busy looking after Saffy. It is good that you are making things for her. She will appreciate that, I know. Well done in your role on the school council. Perhaps you will get to be on a city council one day. I can't imagine you as the nasty scrooge, but the kind scrooge at the end sounds just right.

All the best from New Zealand,
John, Judith, Tim and Hollie.

-

- At [11:00 AM](#), [AAESumner](#) said...

Reuben (and family)

Great to hear about your starring role. Your mum, dad and sister must be very proud. Watch out Bill Murray!

We are all continuing thinking about you and hope things continue to go to plan.

Catch up next week.

All our love

Andy, Anne and Ella

-

SUNDAY, DECEMBER 12, 2004

T +3/ T+4 Still better than we'd been expecting

Hi Everyone

Let's get straight to news of Saff. She's doing fine. She has remained unexpectedly hearty over the past few days, eating well and showing very few signs of discomfort. Today she has developed a couple of problems, an ear infection and what looks like the start of her mouth and throat problems - discovered by an uncharacteristic aversion to marmite!

We have been expecting the ear infection. Saffy gets them regularly, and to last a couple of months without one would be untypical. She has been put on both antibiotics and codeine and remains very chirpy.

One other problem, which has been constant since she's been on the BMT ward, is a very sore bottom, created and continuously exacerbated by the seriously nasty poisons passing through

her body, (sorry for those of you of a squeamish nature, but this site has to be useful to those following us). As any open sore presents an infection risk the BMT team called in the "Tissue Viability Doctors" - more commonly referred to by the nurses on the BMT ward as the nappy doctors (which they love!). We now have anti-fungal cream and that well known favourite vaseline, applied at different times. The improvement to date has been mild.

Saff's hair started to fall out today, apparently she's leaving hair all around the room. We have been warning her about this for quite a while, but rather than let this process drag out, the hospital offer a hairdresser's services to remove it all, and we're keen to opt for this.

I'm writing this from home on Sunday. I should have relieved Mart at the hospital this morning but following a day out with Reuben, (and Mum, Aunty Cath, Aunty Mary and my cousin's daughter Niamh) in London yesterday, I have no voice. Poor Mart has been pushed into a third day of continuous duty, but is stoically claiming he feels just fine.

Must go and tape Sports Review of the Year for Mart - so sorry this is brief.

Take care

Sonja

posted by Saffron Woolley @ [7:03 PM](#)

3 Comments:

- At [9:44 PM](#), [rwoolley](#) said...

Dear Saffy, Martin, Son and Reubs

We are all very happy to hear that Saffy is doing so well. We are very proud of her and of Reuben the actor/politician. Sorry you're not feeling too well at the moment Son, it must be double tough for both of you. Wish you a speedy recovery.

Dex sends his love and agrees with you Reubs, the Incredibles is great, at least up to the bit when Mr. Incredible has a fight with the big robot. At that point we got the fear and had to go home. Oh well, he is only two I suppose. I hope I haven't given him a fear of super heroes.

Looking forward to seeing you next weekend

Lots of love (especially to Saffy)

Rod, Lelah, Dexter and Astrid

- At [12:12 PM](#), Anonymous said...

Re: the little pink bag comments,

1. Toad-in-the-hole consists of sausages cooked in a batter known as Yorkshire Pudding. Yorkshire is England's largest and most beautiful county. The dish is considerably nicer than it sounds!

Hope Emily is doing well and her leg is on the mend.

2. Thanks so much John for a great posting explaining the importance of the work on .

animals with A-Man (sorry if this was you Judith - it reads like John!). To our friends and family outside the ISMRD community, perhaps you can see why it was so important that we flew to Washington back in March. Not only did we get to meet some very eminent scientists (and it was indeed a pleasure to meet Steve Walkley) but we also got to pick the brains of some very dedicated and intelligent parents like John Foreman and Paul Murphy

I hope Tim and Hollie are well, and that you enjoy your festive season - accompanied by some highly palatable NZ red wine.

Lots of love

Sonja

-

- At **7:16 PM**, Anonymous said...

Mac here. Mac here. Mac here. I missed Sports Personality of the Year. i didn't even know it was on. I made a £5 bet with someone at work that Freddy would come second this morning. On shaking he told me he watched it last night. Nice touch. Never paid him, of course. Sorry to hear Saff's off the marmite for a bit- I have deemed as my latest kick on healthy eating and devoured a tub last week. What salt? My hair also falling out but not coming back, alas. Love to you all. Am in London next week if you are around. Now let's see if I can post this only once....

-

MONDAY, DECEMBER 13, 2004

T+3/T+4 - Again, from the hospital

Well, I was wrong about one thing, Saffy hadn't finished her chemo as we'd thought. Yesterday (T+3) the nurse was there, gloved and visored with another black syringe. We had seen methotrexate on her protocol but hadn't actually realised what it was.

It is a small dose of a 4th chemotherapy drug, that is given to inhibit white blood cells called T cells. This is to ward off the particular early GvHD (Graft versus Host Disease) which has been observed in BMT's in the other (more common) storage diseases - the MPS's. Sometimes those kids reject early so the methotrexate works like this;

It targets fast growing cells, including T cells. There are two main groups of white blood cells - neutrophils and lymphocytes. Lymphocytes themselves come in two types - B cells, which create antibodies and T cells (T because they pass through the Thymus gland), they are the front line attackers that target foreign bodies like bacteria and viruses (and hosts).

If the donor marrow is allowed to produce T cells too early, they could attack Saff's cells before the marrow has a chance to start to engraft. The methotrexate knocks them back for a couple of days to give engraftment a chance.

The problem is that methotrexate will stop all new cell production, leaving Saff's body unable to fix itself. At the moment this would mean not being able to get over mucositis (sores or ulcers from mouth through to her little bot). Her bottom also looks like it has second degree burns at the moment and her mouth is getting more sore, so this is an issue.

So T+4 also means the start of 3 days of Folinic Acid Rescue - which sounds a little like thunderbirds to me (F.A.R. Virgil!). This stimulates cell production and will allow Saff's body to recover, or at least not deteriorate.

What else? Well, judging by the liberal dusting of hair on Saff's pillow this morning we will have a bald angel before very long. We are making a real point of showing her the hair and laughing about it - so far she thinks this is hilarious. I'll book an in-room haircut tomorrow. Hopefully we can keep laughing about it all.

Saff also has her first infection, an ear infection. With luck we've caught it early enough and it will be treated aggressively enough with antibiotics (Augmentin) to get her over it swiftly.

Sonja also has our first carers illness, (just a cold) so I'm on a double shift. I've been thinking back and I can't remember ever having this kind of one-on-one time with Saff, in one way its a privilege, she is a quite wonderful person and we are bonding like crazy. One of the best moments was lying on her bed with her (she demanded it) and Saff's snuggling up to me, turning and saying, "Daddy, I happy"

More tomorrow

Martin

TUESDAY, DECEMBER 14, 2004

T +5. Wahey!

Today is the point on this rollercoaster ride when we stopped cranking our way to the top and just began to tip over the edge and get a glimpse of what's below. Saff is getting trouble from the mucositis in her mouth and throat and has stopped eating and barely drinks at all. She lost about 0.4kg since yesterday and her throat is so croaky that she's pretty much stopped talking too. No more 'Daddy, I happy's for a while I suspect.

Her general demeanour is best described as unimpressed and a bit grumpy and, frankly, I can't blame her. I must stress that all the problems she is getting are par for the course and no one here is at all concerned. The symptoms are among those that can be expected at this stage and it is simply a case of dealing with what presents and keeping anything from deteriorating whilst keeping Saff pain and sickness-free until her neutrophil levels start to rise. The anti-nausea meds are working and she has been moved on to oral morphine.

Saff still has raw lesions on her poor little bot. If I'd seen them on her anywhere but here I'd be going out of my mind - and so would she! Just to illustrate that GOSH is not like other hospitals (that I know, anyway) I discussed with the doctor this morning that I wasn't happy with the

treatment for Saff's bottom and within 2 hours, we had a visit from the Tissue Viability Specialist Nurse who made a swift diagnosis and changed us to a different, more appropriate treatment on the spot. It felt like a visit from the Skin Problem Flying Squad. That must have been his Granada Ghia I saw parked outside. Anyway, Saffron's going to love all this open discussion of her bottom when she turns 13....

The best news of the day though, is that on Xmas day they bend the rules and Reuben can come in, meaning the 4 of us can be together. It should be a memorable Xmas, especially if Saff is out of green precautions which might just happen. Keep your fingers crossed for her.

All the best from Team Saffron.

Martin

posted by Saffron Woolley @ **8:46 PM**

2 Comments:

- At **9:56 PM**, [rwoolley](#) said...

This post has been removed by the author.

- At **5:26 PM**, Anonymous said...

Dear all

We've been away for a few days without access to the net (rare I know, but have you ever been to Austria?), and were hoping all was going well for Saff in our absence. So despite the many obstacles which seem to be thrown Saff's way at regular intervals, we are so pleased to hear all is going well, and that you Martin, Son and Reuben are as upbeat and forward-looking as ever.

We're constant loggers-on, and are eager to hear all the news. This blog is the best invention known to man after salad in a bag and the dishwasher.

Thinking of you all.

Love Jon and Cat.

-

WEDNESDAY, DECEMBER 15, 2004

T +6 Time travel

I think this is in fact T+7, so either the toing and froing has queered our pitch, or there is indeed a rip in the time-space continuum.

Seeing Saff today was brilliant and spooky in equal measure. Brilliant because I'm better and was very excited to get to the hospital. Spooky because I knew she would look completely different - but had a suspicion she would look like she did as a baby (Saff was almost entirely bald until she was two), knowing this did not prevent the shock of arriving and finding her asleep on the bed as if, in true Sleeping Beauty fashion, nothing at all had changed in the last two years. It seems we all get to dream.

Martin put paid to my reminiscence by pointing out the astonishing resemblance between Saff's remaining tufty bits and Donald Trump's ginger pate. In his defence, it was a fair call.

Saff's NG tube went in yesterday. She's unimpressed - but it means she can now be given oral medication and sufficient nutrition without too much of a battle. We've been assured that the children quickly get used to the encumbrance, and Erika already feels that this is the case.

Saff's white blood cell counts are now so low (0.02) that they haven't bothered to record the readings for neutrophils and lymphocytes, however tomorrow she is given G-CSF (granulocyte-colony stimulating factor) for the first time-which will help her to fight infection by encouraging neutrophil growth.

We're still probably at least a week away from any early signs of engraftment and the doctors and nurses are keen to point out that this will be a very tough week. We're sure they've said that at the start of every week so far though!

That's your lot I'm afraid

Take care

Sonja

posted by Saffron Woolley @ [10:39 PM](#)

2 Comments:

- At [6:38 AM](#), Anonymous said...

Hello Sonja et al,

I'm becoming addicted to this blogg. I think you are right about the T+7 but you are forgiven. Keep up the news and keep your spirits high. Aren't all our kids just so gorgeous when they are asleep. Love and kisses to Saffy, and big hugs to the rest of the family.

Cheers, John

Petone NZ

- At [11:03 AM](#), Anonymous said...

Hello all,

Saw this joke in the Metro this morning (my journey to work has gone from a great seven minute walk to a miserable 70 minutes on the tube recently so I am reduced to reading rubbish like the Metro). Anyway, made me smile even though it's not very funny:

Q. Why don't clams give Christmas presents?

A. Because they're shellfish.

Am also addicted to this blog - I think it's great, so keep smiling Woolleys and keep typing!

Big hugs and kisses to Reuben and Saffy

Helen xx

-

THURSDAY, DECEMBER 16, 2004



Baby Lucy takes her medicine like a good girl 🗨️

THURSDAY, DECEMBER 16, 2004



Dr Saff will see the next patient now 🗨️

THURSDAY, DECEMBER 16, 2004



Signs of the chemo starting to show, but she's still gorgeous! 🗨️

THURSDAY, DECEMBER 16, 2004



And one of Reuben looking particularly cool on one of his new presents

SATURDAY, DECEMBER 18, 2004

T +8, T + 9, T+ 10. Fox Ward's Star Patient

By T +10, we were expecting to be in the throes of GvHD and fighting infections left, right and centre, but our run of good luck that seems to have followed us this year, continues.* So far.

Saffy has been on great form for the last couple of days. She is on what is regarded as minimal pain relief for this stage (slow-acting morphine), her mucositis and sore bottom have both improved enormously and she is eating and drinking. There is the first possible, tentative, do-we-dare-hope-it sign that her neutrophil count may be on the rise. Her consultant referred to her as his star patient today and it is really hard not to get your hopes up. The fact is, of course, that we can't afford to do that quite yet. Several things could be happening.

One is that the bone marrow Saffy has received has released white cells that have immediately targeted the areas like her throat and bottom and she is getting temporary relief from that. This has no relevance to engraftment of itself. She has also had red blood cell and platelet infusions recently, both of which seem to pick her up. Frankly, who knows. The safest way forward seems to be to do everything we have been doing and quietly keep fingers crossed, while expecting to run out of luck sometime soon. It is almost bound to. 80% of infections come from the body's own flora - bacteria that normally co-exist happily with us, but take their chance to multiply when the immune system is not able to respond to them. And we've always been told that GvHD is a given.

Saffy will be getting her last dose of chemo tomorrow (definitely the last one this time!). It is a final dose of methotrexate to knock the white blood cells back just a little to help counter GvHD and promote engraftment. As before, it will be followed by the Folinic Acid Rescue to start 24hrs later. And I mean 24hrs later to the minute - the accuracy makes a difference, apparently, in terms of cell regeneration cycles. The methotrexate will almost certainly make the mucositis worse, so we could be back to the no eating, no drinking, no talking routine.

If you are getting a taste for these kinds of tales, you might want to check out the site of one of our neighbours - <http://www.teamhartley.co.uk> - it's the site of the family whose four boys all need BMTs and are in with the second son now. Also, if this has made anyone out there think they would like to consider donating marrow, please do take 2 minutes to go to <http://www.anthonynolan.org.uk/ndonors/donor.html> - you really could save someone's life. And you can't say that about most things, can you?

In the meantime, may SuperSaffy's good luck continue and we can't wait to see some of you at Reuben's birthday party on Sunday. Both Sonja and I will be there.

Martin

*I wasn't counting actually hearing about the diagnosis. Obviously that didn't feel lucky at the time, but it's better than not knowing and lots has gone our way since.

TUESDAY, DECEMBER 21, 2004

Saff shows off her Hickman line. The rubber glove is to keep bath water away from the 2 valves that lead into her bloodstream. Hickman lines are a godsend as she has so many IV drugs, but are prone to infections - hence the precautions.

TUESDAY, DECEMBER 21, 2004



You might be able to make out the 2 valves in this picture. Saff is wearing a gauze 'vest' to keep the ends tidy. She is very protective her lines which are officially called 'wiggles'



Taking advantage of not being plugged into any IV meds through the Hickman line or oral meds or food through the NG (nasogastric) tube. I am wearing the obligatory disposable apron - you have to put a new one on every time you enter the room and throw it away as you leave.

T +11, T +12. Looking a gift horse in the mouth.

The blip in Saffy's neutrophil count of 0.12, grew to 0.16. Then 0.31. And today stands at 0.53. Total white blood cell, red blood cell and platelet counts are all rising slowly.

To say that things are going well is an understatement as we should, by the law of averages, have seen infections and GvHD by now. So clearly, we're now worried that our luck cannot possibly last!

Whilst we are trying to enjoy every bit of good news when we get it, we're also trying to remain level-headed about it. For example, acute GvHD can occur up to about T +100, so we're not there yet.

I think the positive thoughts, vibes, prayers and everything else must be working!

Have to run now, I'm afraid, but wanted to share the good news and latest pictures with you all.

Love from the Saffy Squad.

Martin

TUESDAY, DECEMBER 21, 2004



Saff and Baby Lucy saying a really big thank you to everyone for the hundreds of messages, cards and gifts - and all the family seconds that. Thank you so much.

THURSDAY, DECEMBER 23, 2004

T +13, T +14, T +15. It's good news week.

Saff's neutrophil levels continued to rise consistently over the last 2 days. She is now at a whopping 2.91. Amazing to think that just 5 days ago, she was at 0.16 and we were really encouraged by that. This is cracking news and it means that her body can fight off bacterial and fungal infections pretty much on its own now. This is what neutrophils do for a living, but they do not work against viruses, so these are still a concern.

Saff has a dose of a bug called Adenovirus in her gut at the moment. This is not a major concern as it stands and does not even require treatment. If it gets into her blood, though, that is a different story. After the ward round today, I was feeling really bullish and thinking, 'That's it, we're on the home straight now', even though I should have known better.

After the doctors left, I walked out to the kitchen and heard from another parent that their kid had been doing really well – 'All he had was a bit of Adenovirus in his gut; we were getting ready to go home. Now it's suddenly turned up in his blood and we've no idea what will happen or when'. So, back down to earth with a bit of a bump! The GvHD bogeyman is still lurking in the shadows, too, so we'll just carry on enjoying today and see what happens tomorrow.

Speaking of which, Saff has been (or will be) taken off 3 drugs after today's bloods. She will not need 2 anti-fungal medications and an anti-emetic (sickness) drug. She is also moving from IV to oral Aciclovir (prophylactic anti-viral drug). This is exactly what we would have wanted at this stage.

It's just really hard to know how to feel about things and it seems to vary wildly according to whether you are actually with Saff or not. All 3 carers tend to worry less when we're with her. It's strange how walking onto a hospital ward after 2 days away can feel exciting and like coming home.

Speaking of strange experiences, Xmas in hospital is just around the corner now. The ward is decorated, one of the fridges in the BMT (clean) kitchen has been requisitioned for party food, the

nurses station has Xmas tunes playing most of the day and you can feel from the way the nurses are looking forward to it that it's going to be really exciting for the kids. Saff and Reuben will both get presents delivered personally by Santa and as Saff's counts have risen so well, we can all be in her room to open them. Unfortunately, Saffy won't be allowed out of her room, but it should still be brilliant.

There's talk about whether we'll see any celebrities on Xmas day. They had Robbie Williams here one year, you know.....

Have the merriest of Xmasses everyone, here's to a great 2005. Love from the Saffronettes.

Martin

posted by Saffron Woolley @ [10:14 PM](#)

7 Comments:

- At [8:27 AM](#), Anonymous said...

I just wanted to say how delighted I am to hear about latest positive news from the Woolley camp I hope you all have a fantastic Christmas and I hope to catch up with you all in the New Year.

Much love, Jacqui xx

-

- At [10:43 AM](#), Anonymous said...

Great to hear the news about Saffy's neutrophil levels -hope you have a wonderful Christmas (with Robbie Williams)

Lots of love

Simon and Jo

-

- At [11:29 AM](#), Anonymous said...

Mart, Son, Saffy and Reuben (aka Scrooge)

Great to see that things are still going so well. Hope you all have a very merry Xmas.

Best wishes

Mike

-

- At [12:37 PM](#), Anonymous said...

Martin & Family

Have a wonderful Christmas and New Year

Best Wishes

Rosie xx

-

- At [3:38 PM](#), Anonymous said...

Jean here from Chepstow (also from Yorkshire but I now use Auntie Bessie's frozen 4 min. puddings-they're fantastic!) Hope this message reaches you. As a senior citizen my computer skills are not too good. Just to let you know I think you are all great. Tonight we (ie the Choral Soc) are singing carols for the residents of the hotel near Tintern Abbey where C and D had their wedding reception. Tell Saffy that I shall sing "Away in a Manger" especially for her! Christmas blessings and lots of love from Tilly and Hattie's Grandma and Grandpa.

-

- At [7:02 AM](#), Anonymous said...

Hello Saffy and family,

Checking in to wish you all a fantastic christmas day. Our run of bad weather stopped and we had a nice fine day with an outdoor barbecue of lamb, chicken and fish, and lots of other nice goodies too, including a small volume of red wine.

We know it will be different for you there at Great Ormond st hospital but we hope you will all have a great day.

By the time you wake up for your Christmas day ours will be almost over and we will be ready to fall asleep in front of the telly, that is if Timothy is willing to switch off the 12 hours of WWF wrestling DVD he got from Santa.

Special love and hugs to you Saffron on this special day, and hugs to Martin, Sonja and Reuben.

Let us know if any celebrities call in to see you.

From John, Judith Timothy and Hollie.

-

- At [9:52 AM](#), Anonymous said...

Happy Christmas to all of you. Glad to hear Saffy's doing really well and we hope you have a great Christmas, as unconventional as it may be.

Lots of love

Jon and Cat

SATURDAY, DECEMBER 25, 2004

T+16 & T + 17 - IT'S CHRISTMAS!!!!!!!

We're having our fair share of the merry season here, Saff's counts continue to rise, the number of drugs she's taking keeps falling (Morphine and GCSF have been kicked into touch) and she seems really well. Sadly the adenovirus is still alive and kicking in her guts but this hasn't knocked her off course to date and doesn't seem to be troubling her doctors.

Mart and Reubs arrived on the ward with Gaby Roslin in tow this morning – marvellous. I had only just waved goodbye to the chairman and chief exec. of the hospital, wandering amongst the downtrodden to spread a little Christmas cheer. I was starting to feel like a goldfish, but lunchtime sent all visitors scurrying homeward and left the ward to its more long-term inhabitants.

Reub and Saff seemed genuinely pleased to see each other, they needed the smallest of hints to embark in a large bear hug and draw a huge AWWWW from me (and Gaby Roslin!), stood just outside.

The biggest Christmas spread you're ever likely to see was laid out in the playroom for Fox and (close neighbour) Robin wards. Mart has already visited the buffet for two separate meals, and there are rumours it will make an appearance again tomorrow. Santa Claus (the real one you know) arrived on the ward a short while ago. Reub was delighted and Saff terrified as the white bearded jolly man arrived unannounced at her door. She cried so much she was sick and only mildly comforted by the pressie she got. Reub got a very cool transformer weapon (he wants you all to know it's an Armada Dark Sabre).

Thanks a lot for the many messages of good luck and good cheer, we've just been reading them here. A very happy Chrimbo to all of you taking this bumpy journey alongside us – thanks for being there!

Much love

Sonja, Martin, Reub and the Saffster

To the non-Brits:

Gaby Roslin is a minor league UK TV celeb. Actually she was quite nice - and very thin.

posted by Saffron Woolley @ [4:36 PM](#)

5 Comments:

- At [6:54 PM](#), Anonymous said...

Hello La Familia Woolley from the world of fish and chips, sun and ...well....let's say the world island of culture, Tenerife. All is well here. Just me, Mum and Dad. Dad still calling the sun-lounger the lilo, as "vegetarians" they are having venison for dinner, the cat (Billy the Cat) holding the most important place in the family, Dad beginning sentences with "Jeremy, do remember jetty number 8 in Sheerness" and recounting journeys to Poland in 1946. Mum making giant scones. Only six cuppas to date today though. Thinking of you all (including Gabby) . I am just relieved it wasn't Fern cotton on the ward this year. That would have been too much for me to bear. Happy Christmas . Love The Mackinlays

- At **9:16 AM**, Anonymous said...

Glad all is going according to plan. You certainly had a busy day yesterday. Shame about the celeb spot - could have been somebody FAT!!!!

Love from all of us.

The Marrans
xxx

-

- At **11:49 PM**, Anonymous said...

Hello Martin and Sonja, Reuben and her majesty Saffie,
I just read Martin's post tp Paul on the ISMRD'S site and got to chuckling!!

I am ever so delighted to hear how wonderfully well things are progressing with Saffie! I hope the virus works itself right on through with no real difficulty and she keeps going as strong as she's been! I don't want to jinx anything either so I will stop going on about it now but she continues to be in my prayers daily!! Not a day goes by that I don't think about you all and hope things will remain positive. This is a long haul and undoubtedly there will be many bumps but when you have such a strong Queen in charge of things (make no mistake, I was referring to Saffie!) LOL
I think you will all get through this together!! Just keep in mind you have many friends around the world who truly care and pray daily for Saffy's continuing progress!!!

Merry Christmas and God Bless

"Here's to the Healthiest New Year ever!!!!"

Much love and many prayers,
The Haggett's ,
John, Brenda & Zach(ML III)
jhufus@aol.com

-

- At **4:30 PM**, **AAESumner** said...

Its great to hear that things are going well. It sounds like Xmas at Gt Ormond Street is special for the kids.

Gaby Roslin eh! What could be better than that? A real minor celebrity.

Just back from Leicestershire laden with tat, including a huge Barbie castle in lurid pink plastic. Managed to avoid any traditional Xmas family arguments. Maureen sends her best.

Wishing you all continued success and a happy, prosperous and healthy new year.

Hope to see you soon.

Lots of love.

Andy, Anne and Ella.

PS Ella enjoyed Reubens party very much.

-

- At **9:09 PM**, Anonymous said...

Hi Martin, Son, Reuben and Saffy

Have just caught up on a few postings and photos of gorgeous Dr. Saff. Glad to hear you could all spend part of Christmas day together.

The highlight of our Christmas was Rowan opening his stocking and screwing up his eyes with delight at a £2 plastic whistle - the rest of his pressies cast aside for another day.

Nige witnessed a rare win at the Boleyn Ground and wants to know when he's allowed to take Martin and Reuben.

Best wishes - and a happy and healthy New Year.

Kerry, Nige & Rowan

-

WEDNESDAY, DECEMBER 29, 2004

T+18, T+19, T+20. The red hand and the green stool.

Since Xmas, Saff has been exhibiting some of the classic signs of GvHD that we have been expecting. She has been a bit itchy, scratching her head during the night, so we have to massage cream into her little bald head. Her palms have been a little red and itchy, so she has a topical steroid cream. Her poo has been green (are you thinking 'Too much detail' yet?), all of which are classic signs of GvHD.

The fact is that we have been expecting GvHD to hit sometime soon, but keeping everything crossed that Saff is only affected mildly. Without wishing in any way to tempt fate, this seems to be what's happening. She doesn't seem either to be getting any infections. The only problem is that, because it is Xmas, the lab is running at minimum capacity and we are frustrated because we can't get final confirmation on any of these things. Among the top things that we're dying to know are whether there is still adenovirus in her stool (green poo can be a sign of that, as well as of GvHD) and whether it has migrated to her blood. Well, Sonja and I are known for our patience, so we'll wait. Like we have a choice!

There is some more good news. Again, this is a preliminary result due to the lab, xmas etc etc..., but the 'quick' version of another very important test shows that Saff now has 100% donor

marrow. This is exactly what we want (it's what we came in for, after all!). Because this is only a preliminary result, we can't be sure that the actual result would not be 98% donor. We can, however, be sure that it won't be anything like 50%. This wait will be another test of our patience.

Saffron is off more drugs now. She is not getting any pain relief at the moment - so far so good with that one. She is also off G-CSF at the moment (this is the bone marrow production stimulant). This is routinely stopped around now to hold things back a little and hopefully mute GvHD. As a result, her neutrophil counts have fallen back from the heady heights of >5, to around 1.25. This was predicted and is not a cause for worry. Her counts could fall away to nothing again, but at that point they will reintroduce the G-CSF and confidently predict that the counts will come back. This does make us feel like we were justified in keeping our regime as tight as we did when Saffy came off green precautions. Just another dip on the rollercoaster ride!

I just wanted to mention one thing as well. Saffy's playgroup, Leapfrog, have raised over £400 from their Xmas activities on Saffy's behalf and will donate it to Great Ormond Street. That's just another example of what a great pre-school organisation it is. Really professional people, but also enormously caring. When you are in our position, you meet a lot of different people in the local health and educational support network and we hear positive comments about Leapfrog all of the time, which says a lot. Thanks very much to everyone connected to Leapfrog for the collection, GOSH is a really deserving cause. A lot of people know that the hospital is endowed in part by the proceeds from JM Barrie's Peter Pan, but this money will run out in 3 years. Believe me, if you live in the UK and - god forbid - your kids ever need anything from them, you'd want it to have at least as much resource in the future as it does now. If you can nominate your company charities, wonder who to run the marathon for or whatever, please bear it in mind.

While we're on the subject, if you can do anything to support an overseas charity, please make it ISMRD (The International Society for Mannosidosis and Related Diseases) who have helped us so much. They do an incredible amount with next to no money and could achieve a great deal more with more hard currency. If you have any ideas on either score, that would make a real difference.

All the best from Team Saff.

Martin

FRIDAY, DECEMBER 31, 2004

T +21, T +22, T+23. The 'H' word

Sooner or later, there comes a point in the BMT process when most of the drugs are finished, the counts have stabilised (at over the required 0.5 neutrophil count – she's dropped today, but still at 0.95) and there are no infections that need active treatment. At this point there is no further need for hospitalization.

For the first time, we are beginning to hear the 'home' word discussed (by someone other than us, that is). If Saffy can stay free of further infection and we do not get any serious GvHD symptoms, we could be out of here in a week. I have spoken to too many parents who were at

exactly this stage some time ago, and are still here, to take anything for granted, but it's exciting nonetheless.

So, over the next few days, the transition will start from hospital to home care. Saffy has already moved from the intravenous form of Cyclosporin, to the oral version, and her vitamin K will stop when she is discharged, so there is nothing else that regularly needs to go down her Hickman line. The next steps are to train us up to be able to administer her drugs at home and to change to timings of her doses so that we do not have to maintain the current through-the-night schedule.

The possibility of acute GvHD still remains until about day 100, which is a black cloud that will follow us around until mid-March. Acute GvHD comes in 4 levels of intensity. Level 1 is unpleasant and needs careful treatment, level 4 is not something I ever want to experience. No one knows for sure what the determining factors are behind whether you get GvHD. It seems strange to me as a lay-person that 'rejection' can happen so late in the process – after all, if the marrow is going to react against the host, why doesn't it do it straight away? The answer lies somewhere in the complex biochemistry that occurs during engraftment. Needless to say, there's a bit more to it than my Janet'n'John understanding of bone marrow can explain. The fact is we're not out of the woods for another 75-odd days.

So, now we're looking at a new phase in the treatment, there's more to think about. Our house needs to be a lot cleaner than it has been up until now (professional help required), the cats (we have 3) need temporary re-homing, we need to establish workable new routines to cope with having a child at home effectively on yellow precautions. On top of this, we think that this will be the time when we most need to make sure that we are helping Reuben cope with the process. Up to now, he has probably quite enjoyed not having his sister around ("Actually, I find her really annoying most of the time"), and there have been Xmas and birthdays to distract him. Now he will have his sick sister at home, getting lots of attention and monopolising his parents' time to a large extent. That's a lot to deal with when you've just turned 6, so we'll be working hard to help him.

I'm going to leave it there for now. That's enough hedging on my part on a good news day, plus I actually have a bottle of beer to drink on my own for New Year's Eve. I hope Saffy fancies sleeping through, because I might be especially hard to rouse tonight!

Have a great new year everyone. Here's to a really good 2005 for us all.

Cheers!

Martin

5 Comments:

- At [6:00 PM](#), Anonymous said...

Very happy new year to you all.

All our love

The Marrans

xxx

xxxx from Berti and Henri to Saffy and Reuben

-

- At **1:07 AM**, Anonymous said...

Hello Saffy and family,

Happy New Year to all of you. The sound of the "H" word must be great news to all of you. Hope Reuben copes with it OK. I'm sure Saffy will.

Our fingers crossed for things staying good for the next week so you can make that move, and keep a wary eye out for that GvHD thing. It sounds like the model number for a big noisy, smelly, 4x4 off-roader, but I'm sure you wish that is all you had to contend with.

Best wishes from John, Judith, Timothy and Hollie.

Petone, New Zealand.

-

- At **6:54 PM**, Anonymous said...

Dear all

Could it be you're on the home stretch now? Sounds like pretty good news to me. A happy New Year to all of you, and let's hope this year is a lot less stressful than last.

Now, get cleaning.

Lots of love

Jon and Cat

-

- At **9:41 AM**, Anonymous said...

I'm only going anonymous because I don't understand how blogs work. It's Carina here. I've only just discovered your blog so have lots of reading to do to catch up fully. Just wanted you to know that I'm thinking of you and wish you lots of luck and love in the forthcoming weeks. Will write again soon. Carina x

-

- At **2:05 PM**, Anonymous said...

Happy New Year to you all. Good news to read of Saffy's progress, and great news to hear that you can all now think of being reunited at home. Will send celebratory noise in due course. While I am directing positive energy towards Saffy and her recovery, as I read your words, I can really envisage Reuben, and I feel for him. All the luck in the world to you ... Love Is

WEDNESDAY, JANUARY 05, 2005

T +25, T + 26, T +27, T +28. One foot out of the door.

Well, after not letting ourselves get carried away for the last couple of weeks, it seems that we can now definitely (probably) look forward to taking our little girl home on Friday.

There is one more set of test results that we are waiting for (a virus screen). She is not showing any symptoms, so our expectation is that we will get the all-clear. The nagging sense that something has to go wrong sometime might actually be our worst problem at this precise moment.

So we are feverishly making plans for the return home. We are spending money like it is so far out of fashion that we would rather not be seen with it – even by the standard of the last couple of months. Our 3 cats have had to go into temporary accommodation and we have taken the step of having a professional cleaning company come in to get the house ready. Whilst this is considered a positive move rather than strictly necessary, we have taken a no-risk approach so far and who knows whether that has contributed somehow. We certainly don't, so it's no-risk all the way as far as we're concerned. The most important thing to ensure is that the carpets are not left damp as this can encourage fungi.

Sonja is at home with her mum cleaning toys, throwing away junk and generally applying Fox Ward principles. It all makes us feel like this is a new start.

On the medical front, we're learning how to administer Saffy's medicines. They are all going down her naso-gastric tube as she was not happy to take them by mouth. Cyclosporin in particular is very oily and unpleasant to taste and Saffy herself suggested that we use her tube. She will be on Acyclovir, Septrin (aka Co-trimoxazole), Cyclosporin and Penicillin – a much lighter regimen than average as she is doing so well.

Using the NG tube is manageable. You have to draw back a little stomach contents first and test its acidity with indicator paper. This determines that the tube is in the right place and hasn't worked its way into Saff's lungs or elsewhere, which can happen. Then you give the medicines down the tube and follow up with water to flush all the drugs to her stomach and put the stopper back in. Saff is pretty expert herself and 'helps' with much of the procedure.

We will also be getting training on her Hickman line, having a meeting with the senior nurse here and the local team and generally going to school over the next couple of days. More to follow next week at home, I'm sure - we'll let you know after we get there!

It still runs against the grain to say the decision to let us home is finalised, but we hope that the next update or the one after that will be written at home with all 4 of us there. The feeling of relief at having got that far will be huge, albeit mixed with emotion for the families that we leave behind, many of whom are having the kind of very tough times that we were dreading before we came in and, whatever might be around the corner for us, would swap shoes with us if they could.

All the best from Planet Saffron.

Martin

2 Comments:

- At [1:33 AM](#), [Mark Stark](#) said...

Martin and all, congratulations on such great news for the New Year. I know there are many places you would much rather have been this last Christmas and New Year's, so it is fantastic that you'll be going home.

Just a suggestion, we found it quite useful to print out a spreadsheet with the medications and times on it for the week. We'd then check off the medicines after administering them, it was the only way we could keep track during the first few months at home.

Once the medicines taper off, it gets easier.

Let me know if you have any questions, or if we can help in any way.

Best wishes to all,
Mark and Kathleen

-

- At [1:36 AM](#), [Mark Stark](#) said...

Martin and all, congratulations on such great news for the New Year. I know there are many places you would much rather have been this last Christmas and New Year's, so it is fantastic that you'll be going home.

Just a suggestion, we found it quite useful to print out a spreadsheet with the medications and times on it for the week. We'd then check off the medicines after administering them, it was the only way we could keep track during the first few months at home.

Once the medicines taper off, it gets easier.

Let me know if you have any questions, or if we can help in any way.

Best wishes to all,
Mark and Kathleen

-

SUNDAY, JANUARY 09, 2005

T29, T30, T31, T32. Home Sweet Home.

After what seems like the longest time one minute, and an amazingly brief time the next, we are now at home. It is an enormous relief to be here. Of course, getting here was the usual Woolley family mini-epic.

At one point I thought Son was going to suffer some mental trauma induced by the scale of the clean-up operation at home whilst I was doing the last shift at Fox Ward. In typical fashion, she set herself very high standards and then met them all, with the result that the house was not only as hygienic as it has ever been by a comfortable margin, but Saffy's room was a pink paradise

covered in (left-over Xmas) presents when she arrived. The look on her little face was priceless. Son will freely admit that she could not have got close to getting everything done without the drive, determination and elbow grease of Margaret, her mum, who came down for 3 days from Worcestershire, just to help out.

Saffy is now spending her time going and finding all her favourite things at home and in between time just grinning. She seems really well 'in herself', as they say. Going home seems to have given her another boost, as did the red blood cell transfusion and the white-blood-cell-boosting G-CSF infusion that she had just before leaving hospital. Both counts were a little on the low side and the doctors wanted to give her body a little extra help to cope with the outside world. Or the limited part of it that she will see from inside the house.

We have set up a M*A*S*H*-style field hospital in Palmers Green with syringes, medicines and other paraphernalia around the house. So far, managing the medications has been fine - Son and I supervise each other to make sure we're putting everything we were told into practice. Keeping the house hygienic is a challenge - Reuben and Saffy managed to wee on or near each other and on the floor within about 1 1/2 hours of getting home.

We have a way to go yet - it's only been 48 hours since we got her home - so, we do not want to take anything for granted, but while the hospital stage that we have just completed is still fresh in our minds, we wanted to note a few things that felt like they had worked for us for anyone reading this in the future preparing for their own child's BMT. This all comes with a health warning because neither we nor anybody else knows anything for certain, but for what it's worth, here goes:

Things Outside Our Control:

We had a good match - even though it was an unrelated donor (10 /10 tissue match, later further refined to 11 /12).

Our donor was physically large, meaning that we could get a lot of marrow - the large volume may have had something to do with how quickly Saffy started showing neutrophils post-transplant.

We had a male donor. Male to female transplants are the most desirable. Frankly, we can't remember what the reason is for this, but the outcome tends to be less severe GvHD.

The support we receive from our friends, family and employers is as much as anyone could wish. Seeing other families cope with less has brought that home. Because of this we have been able to devote ourselves to Saffy and Reuben during this time. If we get through this whole process relatively unscathed, this will be part of the reason why.

The team at GOSH. They are absolutely brilliant. Just brilliant.

Things Within Our Control:

All of the things above (plus luck) probably account for over 90% of success in BMTs. Through what we did we tried to squeeze in another few percent difference. What we did was:

Set very high standards of hygiene. We took the cleanliness rules as gospel and wherever we were told 'do this 3 or 4 times a day' we tried to make sure we did it 4 times, not 3. It is quite possible that either Sonja, Erika or I will have obsessive-compulsive disorder in later life.

Think like a germ or 'If I were a rotovirus, where would I hide'. When you're not doing anything else, look around and find spots like door handles, the tops of high shelves etc and get happy with the alcohol wipes.

Find the nurses who have been on the ward the longest and quiz them. Find out everything you can. Nurses with a strict demeanour are the best for this. We loved the strict nurses, because they weren't afraid of hurting feelings, they just said what they thought was best - for example good ways of separating gloves used for different functions to different parts of the room. Sounds anal. It is. But these nurses really knew their stuff and it makes really good sense.

Talk to everyone. The housekeeper can tell you really useful things like where it's safe to get bread in the middle of the night when the frozen, individually-wrapped portions have run out in the special BMT kitchen and you have a girl suddenly demanding toast after 36 hours. The cleaners are sometimes the same ones who prepare rooms for green precautions - they know more about killing germs in your room than you do and they're full of tips.

Be strict with each other. The 3 carers helped by policing each other - it is really easy to forget instructions when you hear so many, so it helped that we reminded each other.

Take short shifts if you can - 2 nights was our ideal when possible. There is a lot to do, it is stressful in different ways and the nights are always broken, so being able to rotate carers regularly is a good idea.

Write everything down. Take notes when you are learning about things. Also, write everything down during the day. You won't be able to keep a note of when she drank what, which cream you applied when, how many times you did the oral care routine and everything else, all in your head.

As well as all of this, try to make the experience as much fun for your child as possible. This might sound strange, but when you're a kid, there can be fun in anything. Saffy enjoyed watching her old favourite videos as much as she wanted (I can kill at the mere sound of the Balamory theme tune now), getting lots of new sticker books and even playing the 'Who can sellotape up most hair from the pillowcase' - a game that you can only play under very few circumstances! Most of all, stay happy yourself so you create a positive atmosphere.

None of this is rocket science. More an extreme form of common sense. Don't feel too daunted - we learned everything from friends with previous experience, but mainly from the staff at the hospital who are used to guiding families through this, and who could, if necessary, take your child from beginning to end of the process without you.

Anyway, enough top tips. We'll let you know how well things are going at home soon.

All the best from a very happy Team Saff.

Martin

PS. I also matched Son's 'Raspberry Nipple' cock-up, when we were getting the last few things together before leaving. As I tried to reach a particularly elusive get well card from a high shelf, I over-reached, but managed to grab it. As I turned to leave the room I was almost knocked down by the charge of nurses and doctors arriving within seconds from other patients' rooms. Obviously in over-reaching I had leant on the crash alarm placed out of the way high on the wall and they were arriving to save somebody's life. I was left in no doubt what a medical faux pas I'd made as I walked past that same team on our way out....

posted by Saffron Woolley @ [9:04 PM](#)

5 Comments:

- At [2:39 AM](#), Anonymous said...

Congratulations Saffron and team for making it home at last.
Here's hoping for a great grafting time in the weeks ahead.
Love and hugs from John, Judith, Timothy and Hollie

-

- At [5:28 PM](#), Anonymous said...

Great to hear that you're all back home. Hope everything continues to go so well.

All the best

Mike

PS Son - you can do a few hours a week cleaning my flat if you fancy it.

-

- At [9:22 AM](#), Anonymous said...

It's fantastic news that you're home. Isn't it an awesome feeling. Congratulations on all the hard work.
Luv from Sonia, John, Lewis & Ryan Dernie in NZ.

-

- At [8:23 AM](#), Anonymous said...

Great to hear that you're home. Having spent a lot of time myself at GOSH (nothing in comparison), I can vouch for their amazing kindness, but there's nothing like home. Will follow your progress with interest.
Carina x

-

- At **10:05 PM**, Anonymous said...

Dear Martin, Sonja, Reuben and Saffy!

HHhhhoorrrraayyyyyy!!!!!!!

I'm so delighted to hear you are home and that Saffy's doing so well!

You have remained so strong as a team and I'm sure you will continue.

John and I will keep you all in our prayers that Saffy can continue grafting straight through the next 90 some odd days!!!

You all are never far from our thoughts and I can't wait to read your updates!!

Keep up the good work Saffie and team!!!

Love and prayers, and many (((hugs)))

John, Brenda and Zach Haggett, NY / USA

-

TUESDAY, JANUARY 25, 2005

Deja vu (all over again) Up to T+48

So, here I am at home again with Reuben whilst Son is in hospital with Saff. It's all strangely familiar. Don't be concerned, Saffy is in hospital for her flebogamma (immunoglobulin), which she needs intravenously every 3 weeks. She was also anaemic (Hb of 4.2), so she needed a blood transfusion. She is at our local hospital, who now share her care with GOSH, and not every paediatric ward works with Fox ward's speed, so it has turned into an overnight visit. Saff had been complaining of headaches and tummy aches, but had not developed a fever - we are hoping that she will get the boost from the red blood cell transfusion that she did before. No one at Chase Farm (our local) or GOSH is too concerned - sometimes new marrow just does not produce enough on its own at this stage and some extra is needed. To be on the safe side, they are going to run some tests overnight to see why the red blood cell count is dropping. The 3 usual reasons would be an infection (no sign of this), blood leaking somewhere (ditto) or haemolysing, which, as far as we understand, is the body breaking down red blood cells too readily. If it is this is happening, it leaves markers that can be traced and we will find results out tomorrow.

May I apologise that it has been such a long time since you have heard from us. No excuses, we should have let you know before now what has been happening and I hope that not everyone has given up looking out for updates.

I have gone back to work now, so the feeling of limbo between leaving hospital and getting back to a closer approximation of normal life is starting to disappear. We have set up our home and our routines and support networks pretty well. Medication sets the tempo for life. As suggested by experienced friends, we made spreadsheets of all of Saffy's medications, setting out times and doses. Even though Saffy is on the minimum medication for this stage, it is still remarkably easy to make a slight slip and having it all on paper makes a that a lot easier.

As I mentioned, her care is split between GOS and Chase Farm hospitals. Chase Farm send a community nurse every week to change dressings weekly and new NG tube monthly. They also take bloods on which they can do the more usual tests for cell counts and biochemistry. On our clinic visits to GOSH, they take blood for the more specialised tests, which includes cyclosporin

levels. Cyclosporin is one of the key drugs at this stage. Its job is to mute any GvHD, which is important, but in doing this, it also suppresses the lymphocyte levels. This is partly why Saff's immune system is still not working at anything like full capacity. The ideal level is around 150 and, according to the blood test results, her dose is adjusted to try and keep it thereabouts. The theory is that she gets minimal GvHD and they will then gradually bring the dose down and tail it off. Fingers crossed, as usual. That approach has worked until now, so we're not changing.

Day-to-day, Saff is still delighted to be at home. She is starting to show signs of feeling the limitations of her life at the moment and of reacting to all the attention she has inevitably got more used to. This is manifesting in demanding more attention, thinking that she is going to be going to school with Reuben the next day and so on. This is really not much, and she is still far more likely to get excited about getting a visit from someone than worrying about what she's not getting. Her glass is always half-full.

Speaking of visits, we can have 3 or 4 people round to the house at one time, so long as everyone is healthy (no coughs or sneezes, no runny noses, no diarrhoea is the mantra). Other people we know have had more stringent rules on being discharged - we can only assume that this is down to how well Saff is, relatively speaking. This makes a lot of difference as it allows everyone - Sonja & Saff in particular - a break from being in each other's company exclusively, as well as giving friends and family the chance to see Saff for the first time in several weeks. Saffy is also allowed out of the house, but only where she can maintain some distance from other people. At this time of year, that means that if we wrap her up in about a dozen layers, we can get out to a park for a run around as they are very sparsely populated. Shops, restaurants, indoor play areas etc are all no-go areas.

I'll leave it there for now, but we will - we promise! - put another entry up in a day or two to let you know what happened with the blood tests. Radio Saffron is very definitely back on the air.

All the best

Martin

FRIDAY, FEBRUARY 04, 2005

Music to our ears.....to + 58

I'm writing this entry from our local hospital as Saff has a blood "top-up". Everything continues to go really well.

We were at GOSH yesterday and saw Paul Veys - he's the "grand fromage" of the BMT unit and was really delighted with Saff's progress to date. He confirmed his belief that Saff's falling Hb levels are caused by immature marrow saying the red blood cells are typically the last to stabilise post transplant. He did however test another couple of possibilities to exclude them. The first is caused by the Cyclosporin attacking the cells, they're left looking like they've had a huge bite taken from them. Treatment is simply to withdraw the Cyclosporin. The second could be Saffy's

original B cells attacking the donor's AB cells - this can occur because B blood cells contain A antibodies. If this is occurring they simply wait until all Saffy's residual B cells disappear and she takes on the donor's AB blood group.

Today's transfusion is to boost her levels closer to normal and then watch to see how long it takes to fall. Paul Veys says even if this process (i.e. transfusion followed by falling Hb) continues for a number of months he will remain unconcerned.

We left GOSH and travelled to the local hospital for the transfusion (obviously!). As Saffy had had blood taken 7 times in the last 10 days it stands to reason more would need to be taken to establish what blood should be ordered!! We arrived at 2.30pm, had blood taken at 3pm, and were told at 6.45pm that although the blood had arrived it was the wrong type. Words failed me - well actually they didn't but I think the nurse who had to tell me this wished they had.

So here we are this morning - I'm considerably calmer now the process is almost over. We're going home so Saffy and her nursery teacher Natalie can have fun. Tuesday's "lesson" involved a fantastic shaving foam fight. It's a pleasure to hear them at work!

More soon

Sonja

P.S. All blood results for the tests above were negative - brilliant

posted by Saffron Woolley @ [8:48 AM](#)

5 Comments:

- At [11:17 AM](#), [Bristol bunch](#) said...

Glad to hear the continued good news - we keep a regular eye on the Blog. Have wonderful image of Sonja losing her world famous patience with some poor unfortunate nurse....you need to let off steam and better them than Mart!

Keep smiling

H, H, & E

-

- At [4:29 AM](#), Anonymous said...

Hello Saffy,

So pleased things are still good. Tell your dad the England team did really well at the Rugby Sevens in Wellington last weekend, beating those great rugby nations Niue, Canada and Samoa, but going down to Fiji and SCOTLAND. Poor dears. Tim and I were there having a great time. Tell Martin, in case he missed the news, that WALES also dealt to the England rugby team on the weekend. What is all that about World

Champions? Must mean "used-to-be". Martin might need some cheering up, so make him laugh. Best wishes,

John
Petone, N.Z.

-

- At **1:32 PM**, Anonymous said...

Hi Martin, Sonja,

Glad to hear things are going well overall. Pam and I have been thinking of you all, and keeping our fingers crossed.

All the best,

Sumant

-

- At **11:02 PM**, Anonymous said...

Hello Woolley Family,

Very glad to know all is going well. Have tried to contact Erika and Bill for a personal update without success. Away now for three weeks. Will keep rooting for you,

Mary and John Samuel

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- At **4:28 PM**, Anonymous said...

Dear Saffy, I was really chuffed to read your news , especially the bit about how much of your treatment is now either at home or closer to home. We are looking forward to seeing you soon, but as there hasn't been a frost to get rid of all the winter lurgies, we've had a few hangers-on testing us all over the past week. My biggest challenge was trying to persuade Maya and LB to use pull-ups. I failed. Roll on Easter, better weather and all the joys of spring. Love Is, Andy, Maya and LB Cooper

WEDNESDAY, MARCH 02, 2005

More of the same..... to +84

Sorry for the big gap - but it's getting harder to find things to tell you about here in Camp Lucky. Saffy remains happy and healthy. As yet we have no solution to the continued problem with the red cell count but all medical personnel remain remarkably calm and we are doing our best to mimic that state. In fairness, it's not that hard because Saff looks and acts like a girl that's never been ill in her life.

On our most recent trip to GOSH - last week - the BMT consultant said all that worried him about Saff's tumbling Hb levels was his inability to pinpoint the precise cause. He thought her residual A antibodies attacking the donor AB marrow was the most likely explanation. He felt the immature marrow theory would also lead to reduced white cell counts and this hasn't been the case. What is an undisputed fact is that a significant number of post-transplant children suffer from this problem, and it almost invariably corrects itself, regardless of why it exists in the first place! She hasn't had a transfusion for over two weeks now and although her Hb level is low it's taken longer to fall than usual - so fingers crossed.

I have to confess that all this staying at home has given me an acute attack of cabin fever - I find myself looking forward to shopping trips - and I don't even mean for clothes! Still, all in a good cause.

Reuben, Pokemon master supreme, top Hogwart's wizard and family Top Trumps champion, continues life unconcerned by the endless medical interventions needed for him and his little sister - thank goodness.

Thanks to all of you still following our fortunate progress - I'll try and update you a little sooner next time.

Take care

Sonja

posted by Saffron Woolley @ **9:26 AM**

1 Comments:

- At **8:27 PM**, Anonymous said...

Hello Sonja and team,

I'm committed to the idea that no news is good news, so despite my regular checks of your site, I've been happy to see few postings of late. We are all so pleased things are going so well for Saffy.

Perhaps you could cope with cabin fever by writing a book on "The joys of grocery shopping". It really is one of life's great pleasures. I found it good for quality time with the kids, and I bet you could really appreciate the opportunity to discover the fun side of it!!!!

Love to all of you,
John and family
Petone, NZ

THURSDAY, MARCH 31, 2005

Last lap? To T+113

Is there anybody out there? Couldn't blame you if you had given up on us after so long without an entry, but as several have mentioned, no news tends to be good news and that has been the case with us.

We are at the point now where Saffy's last immunosuppressant drug (Cyclosporin, aka Neoral) is being eliminated by stages. We are down to 0.3 ml twice a day, from a high of 0.55. This drug suppresses any GvHD, so we can expect to see some sign of it as the dose comes down. So far, all we have seen is the intermittent rash we saw in hospital, which appears and disappears after a few minutes. If we can get away with nothing more than that, it will be a miracle, but then, everything has been a bit miraculous so far.

The issues we have been contending with so far have been different to those we were expecting, as Saff has been so well. With me going back to work, Sonja has been pretty much stuck in the house as Saff is not able to be around groups of people. For someone who has been used to working full time, that is a hard transition to make and it has taken quite some getting used to. For my part, I have been working full time for quite a while and taking over on duty from Sonja at weekends. The practical result of all of this is that it takes quite a bit of planning to do anything outside the usual routines and time out of the house for Sonja and me together, or for us to take Reuben out. Don't get me wrong, we still thank our lucky stars each day, it's just that we are not living what you could call an everyday life yet. Between Reuben's diabetic care and Saff's medication there are around 8 entries in our daily medical diary, when they are both at home, so it is a bit of a stop-start existence.

It has been a great help to us all that friends and family are really putting themselves out for us and the messages we continue to get from friends through ISMRD and GOSH reminds us that we're all getting through with each other's support.

Have to go now - it's bathtime and I've got some Hickman lines to wrap in clingfilm....

All the best

Martin

TUESDAY, JULY 05, 2005

T+208. A trip down memory lane...

I can't believe that it's been THAT long since we put an entry up. I mean, I know it was a long time, but....

This is for whoever's reading this in the future, as there's no one who can still be checking for an entry after 95 days of not seeing one.

It seems like a lifetime ago that those entries were coming thick and fast. Since then, it's been like an uneasy limbo. I'm working, Sonja can't think about getting out to work and Saffy can't think

about playgroup as every last aspect of our near future is governed by those weekly lymphocyte readings. Saff is doing really well, but it seems that lymphocytes after alpha-mannosidosis BMTs just don't come back quickly. About the same speed as a glacier. We know how lucky we are - we really do - but it just isn't possible to feel lucky all day for weeks and weeks.

Among other things, this has been a voyage of self-discovery for us, and we now know that our patience has limits and that we are better suited temperamentally to the parts of the process that involve immediate action.

So, when Saff developed her first line infection three days ago, we were able to get back to what we do best and react to an evolving situation. To get to 205 days past transplant without having a line infection may be a record - many of our contemporaries in Fox Ward needed all their fingers and toes to count theirs after much less time. Anyway, we hadn't seen one, so it came as something of a shock...

We were actually at the UK MPS Society conference, picking up hints on trigger finger (which Saff is just starting to get) and biomarkers (biochemical indicators of disease), when we came out of the last session, turned on the mobile phones and got a message from my dad that Saff had a temperature of 39.5 and was on her way into hospital (GOSH organised everything so that she could go into our local hospital). By the time Son and I arrived (in about an hour Northampton - London, hope we didn't pass any speed cameras) Saff was over 40 degrees, fitting, frothing at the mouth and apparently awake, yet unconscious and unresponsive. Pretty scary, but bringing her temperature down brought her back - suggesting that this had been a febrile convulsion.

GOSH took no chances and ordered a head CT scan to check for abscesses on the brain, a lumbar puncture to check for meningitis and a chest x-ray. Saff's temperature was 'spiking' - coming down with paracetamol and ibuprofen, then shooting up as it wore off. She was put on antibiotics to cover all major potential problems - particularly meningitis. Her lymphocytes dropped to 0.15 from around 0.8 a few days before. She was clearly ill, but, as ever, cheerful as she could be. One by one, the really nasty things got ruled out, thank goodness.

Anyway, within a day the hospital had identified Staphylococcus and Saff had another antibiotic added to her regime. We already 'knew' it was from her line as she had gone downhill within minutes of her Hickman lines being flushed - a sign we'd been told to look out for as the bugs get flushed into her bloodstream from the line where they were sitting, causing the sudden, dramatic deterioration we had witnessed. Tests confirmed this later and Saff is doing much, much better now the bacterial infection is being accurately and aggressively treated. We expect her out in a few days - maybe sooner.

Being back in a hospital room and going through all of the old routines brought it all back (probably why I did this blog, too, I suppose). If the infection doesn't get cleared from her line, it may well be taken out a few weeks ahead of schedule and GOSH think her counts will bounce back once she is better.

The medical staff at both hospitals were really great - professional and caring and made us part of the team immediately and a great 'catch' by Saff's grandparents who did exactly the right thing by calling them as soon as they got concerned.

If all this excitement keeps up, I'll be putting another blog up soon - great to hear from messages that it has found some people in similar positions already and been of some use.

All the best for now

Martin

THURSDAY, DECEMBER 22, 2005

T+365. Signing Off.

Well, one year on - almost to the day - and we have been signed off by the BMT unit.

When we started this process by Saffy being recommended for a transplant and then starting the search for a donor about 18 months ago, we had little idea what we were letting ourselves in for. We had only an outline idea of what we were letting our little girl in for.

We were convinced that a BMT was our only hope of giving Saff a fighting chance of something like a 'normal' life. The odds were good - around 90% of BMT kids survive the procedure and many of the 10% are those poor kids whose conditions (including cancers and auto-immune diseases) had weakened them already.

Over the weeks however, it's fair to say that the 10% started to haunt us. Living with the prospect that the worst might actually happen had several effects on us. We prepared like mad - emotionally, practically - we thought about what Saffy would need, what her brother Reuben would need. Who could help with what, who would need what. In all of these preparations our ISMRD friends were immensely helpful and knowledgeable. We made arrangements about work and childcare and my mother agreed to be our 3rd carer. We noticed somewhere along the way that we had stopped being interested in some of the things we used to do that somehow just didn't seem very interesting any more. We probably drank too much.

And then we were in GOSH and life had its own momentum and intensity. We worked at everything we could, learning as much as we could from everyone - medical and other staff at the hospital, other parents, other kids. I won't forget some of the most profound lessons. In a sense, this was the easiest part - it was intense, there was always something that needed doing. We work in deadline-driven jobs so this part played to our strengths. It was all about Saffy (and Reuben) and we did not have too much time to sit and think. Doing the blog every night was therapeutic too.

But, above all, we were very, very lucky. We were well-prepared and expecting infections, rejection, extreme reactions to medication. In the event, none of these turned out to be as bad as we had feared and Saffy was an absolute delight. Her anxieties were there. We saw them for example in her developing a fear of clowns (called coulrophobia, if you're interested) whilst in hospital - but she found the good in every day and every situation.

And now, here we are. BMT officially over. It has been the single defining episode of our lives so far. For Saffy, of course, it is her chance to develop. Her alpha -mannosidase level is 77 on a scale that runs to 520, which puts her at the low end of carrier level. This could go up, but to be honest, our doctors have no idea. What it means to her development will unfold in the years to come. Her doctors, teachers, psychologists, occupational therapist, speech therapists, ophthalmologist, spinal consultant, audiologist, dentist and the rest of the team will monitor and help her.

As for the rest of us, Reuben carries on pretty much as before. We consider this a victory. I am not the same, largely through learning something about myself and my priorities in life. I won't bore you with all of that. Sonja having given up her job to care for our kids through Saffy's BMT is applying to become a teacher. The plan is that this gives her more time to spend with the kids and, although she has not said so, I suspect the opportunity to help and nurture children in some way has more of a magnetic pull now than maybe it would have done before. Getting onto a course will be a new challenge, so fingers crossed.

Before we sign off and complete this blog, which we really hope will be of some help to people starting on the same road in the future, we want to thank all our friends and family - both those we knew before and those we have discovered during the last year and a half. Because things have turned out well, what has happened is that many of our relationships have been strengthened and we have found others who have entered the Woolley Family Hall Of Fame. This isn't Oscar night and I am no Gwyneth Paltrow, so we'll leave it there, but because of the people around us, we believe we are luckier than most.

Thank you.

All our love,

Martin, Sonja, Reuben and Saffron Woolley.

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