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18 September 2007

Appraisal of drugs for the treatment of pulmonary arterial hypertension

One evening in February 1999, when I was only 23 weeks pregnant with our third child, I coughed and there was a very unpleasant taste in my mouth. On coughing again, I realised that I had, in fact, brought up blood. I suddenly panicked: I'd never been ill before, other than your normal coughs and colds, but this was serious.

After phoning the emergency doctor I was advised to go straight to our local A&E department. It was now probably about 11.00 or 12.00 at night and our two children – Natasha, barely 14 years, and Daniel, 10 – were in bed fast asleep. We woke Natasha and told her that I was feeling poorly and had to go to the hospital, but not to worry because we would be back as soon as possible. Little did we know that I wouldn't be back home for a further eight weeks…

I was diagnosed with primary pulmonary hypertension within the week. It was described to me as 'untreatable, incurable and terminal!'
At one point it was suggested that I might have to terminate the pregnancy…well I already had two children. I made it very clear that I was not prepared to have a termination. So further tests had to proceed with caution.

My pulmonary artery pressure (mPAP) was rising constantly, for example a persons mPAP at rest is normally 14mmHg, mine was over 70mmHg. All due to the extra pressure your body is under during pregnancy.

It was agreed that the best way forward was to have an elective caesarean. The date would depend on the health of the baby and how my heart was standing up to the rising pressures in my pulmonary arteries. Once these had risen to above 130mmHg, it was time to deliver. By this time I'd been in hospital over six weeks without being allowed home. Dan used to bring the children in daily and coped pretty well with the frequent trips to and from the hospital. Most days he would make two journeys. He hadn't had to look after the children like this before; I used work and make all the childcare arrangements. He found things a little tricky at times.

I was allowed home the weekend before the planned caesarean to 'put my house in order', as I was told that I had a less than 50% chance of surviving the birth. Records showed (at the time) there had only been 20 cases of the mother surviving the birth, or so we were told. I was determined to be the 21st!

After the delivery of our tiny baby, Maddison (weighing 1.1kg), I spent three days in intensive care and then a further week on a high-dependency ward. I felt awful. I was on oxygen 24/7, as my saturations would fall as soon as I moved any part of my body. If I thought I didn't feel too bad before I had my baby, now I was in shock. I was breathless, had irregular heartbeats and chest pain all the time. Not to mention the baby blues. It was nearly a week before I got to see Maddison. She was so tiny and fragile, but I knew that, if she could make it, then I would too.

I was allowed out of hospital towards the end of June, in a wheelchair, and told that I would be referred to the Queen Elizabeth Hospital in Birmingham for a transplant assessment in October, six months after giving birth to Maddison.

The first twelve months after being diagnosed were the worst of my entire life. I had a very low period after being discharged from hospital: I was crying a lot and thinking about dying. Looking back, I now feel I allowed myself to grieve, to try and get it out of my system, because I knew I had a fight ahead of me. It wasn't easy, but I've never had things easy, anyway. So the fight began.

Before I was diagnosed I had always worked. I started my working life as a florist, after having my daughter, Natasha, at eighteen. After the birth of our second child, Daniel (the 'devil child from hell'), I retrained as a printer and after successfully managing my own department moved into the publishing side of things. I was working in the publishing department of a government education agency to promote the use of ICT in education when I was diagnosed with PH. Everybody there was very supportive and, as my colleagues/friends had unlimited access to the Internet, they turned into my researchers, finding out all the background on PH and who was doing what and where in the field. A lot of the information that was available came from the US; the statistics and figures did not paint a positive picture. The prognosis was very grim. However, I wasn't going to let this deter me. I wasn't happy to sit at home and wait to die so, against the advice of my cardiologist, started a staged return to work. If I had to sit anywhere, then it would be behind my computer doing something worth while and holding onto every bit of reality I could.

I was very uncomfortable with the survival rates of lung transplantation. Even if I had been lucky enough to survive the first three years, it would be very doubtful that I'd be here today. After I'd been for my first transplant assessment there was no way I was going back the following month for another. I wanted to be referred to a professor based in Sheffield that I'd found out about on the Internet.

Eventually I was referred on and I saw Professor Higenbottam in November. During the initial consultation we discussed what medication was available and he informed me of two drug trials that were being started in the New Year – one on nebulised iloprost (inhaled) and the other, slightly later in the year, on UT15 (subcutaneous infusion).

After the baseline tests were completed –(lung function, walking test and right heart catheter) plus the paperwork, the trail began. It was quite difficult at times because you were expected to use the nebuliser nine times a day, sometimes for 15–20 minutes at a time. It could be exhausting. The technology of the

delivery of the medication has taken eight years to refine. After completing the trial, I was allowed to continue the therapy for the next five years or so at the expense of the drug company.

Without the medication I'm sure that I wouldn't be here today. It allowed me to build up my strength gradually in order to get through the day. I have faced up to the fact that I cannot do all the physical things that I would have taken for granted before my illness, for example climbing a flight of stairs, walking up a slight incline, doing the housework (which isn't so bad!), but it's the times when I haven't got enough energy to lift a limb or wash my hair that sometimes gets me down. If I stay up too late to finish something or perhaps do a little too much around the house, then I know about it the next day. It's pretty counterproductive really because I don't get any further.

The immediate family have been fantastic on the whole. My husband had to give up his job as a carpenter to become my carer, not to mention caring for the children, who are now 23, 19 and eight. In fact our eldest daughter has made us grandparents and we have a beautiful granddaughter called Millie-May, who will be three next May. I never imagined I would ever see grandchildren.

Being a 'professional patient', attending appointments every three months in Sheffield and every six months in Coventry, we have met a lot of other people that have also been diagnosed with PH. One of my very first memories was of being introduced to a young man, barely twenty, who had started to become severely breathless and lethargic, only to be accused of trying to skive off work and being called lazy. He could hardly move off his coach. It took him some time to be diagnosed, but eventually was seen by the specialist centre in Sheffield. Because his PH was so severe he immediately had a Hickman line inserted. One of the problems with having an IV is infection and this chap was no exception, he was being treated for an infection in his line. It's very common for this to happen, but now there are many more options available to the prescribing physician, before having to take such drastic steps.
Patients can benefit from less invasive therapies, but using the same technologies, such as inhaled medication and oral medication.

On my return to the hospital, three months later, I enquired about the young man. Sadly he had died. I have met a few patients that have lost their lives to this debilitating disease, but many more that are benefiting from the medication available today.

In 2004 my specialist introduced a second therapy to my daily ragime and, after waiting nine months for funding, I am now on Ventavis (nebulised iloprost); with a fantastic new delivery system (well done, Respironics!), and sildenafil. Both therapies relax the walls of the blood vessels and allow more oxygenated blood to flow through the lungs. After three months of taking the sildenafil I felt a huge improvement in my exercise tolerance. By this I don't mean going to the gym or jogging round the block, just getting up and getting Maddison off to school or coping with the problems of having a teenager with attention deficit hyperactivity disorder (ADHD). So now I'm on Ventavis, sildenafil, oxygen therapy during the night (and sometimes during the day if I've overdone it…), warfarin to maintain an INR of 2–3, mesalazine because the iloprost has given me colitis and esomeprazole because the sildenafil has caused reflux. A small price to pay for

being given a chance to continue to raise my family and contribute to society. My family have adapted well to my PH and know that it will always take us longer to do things and plan holidays with military precision, but I'm sure they would rather that than not have me around at all.

I have also been a trustee for the PHA UK since 2002: contributing to the work the charity undertakes for example; campaigning to tackle serious issues such as funding for PH services, raising awareness of the disease and the problems our patient community face. Without addressing these issues some of our patients will fail to receive the life-saving treatments and support that they need.

After some serious soul searching, I decided to hand in my notice at work, a job that I dearly loved, to do something that I have always wanted: go to university. I could have carried on working, but I found that when I returned home in the evenings I was very tired and had very little energy for the children or even a conversation with my husband. So I said goodbye to my colleagues and enrolled on a foundation course in art. I completed the year with distinctions and went on to start a degree. I've had my ups and downs with health and family issues and have settled on doing the course part-time with a view to teaching teenagers with learning or behavioural problems. I may not ever be in a position to achieve my goal, but I'll give it my best shot with the benefit of the therapies we have today and those that are being developed for the future.

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