

## Living with ARVC

Having had my first ICD fitted by Dr Bennett in 2003 and being somewhat reserved about discussing my condition with people other than health professionals, I felt it was about time that I finally put pen to paper.

I have ARVC (Arrhythmogenic Right Ventricular Cardiomyopathy) which is believed to be an inherited heart muscle disorder where damaged muscle, over time, becomes replaced by scar tissue and fat. Symptoms vary, but in my case only surfaced when I reached the grand age of forty! At that time I was a serving officer in Cheshire Police and had been physically fit and active since a child. I played competitive sport such as badminton and football but subsequently devoted my energies to becoming a runner. I was very keen, trained about five times a week (where duties permitted) and represented the force at various events as well as at my local club.

In 1999 I began to experience palpitations and occasional breathlessness, sometimes during running, others whilst at relaxation. Tests showed nothing untoward and the problem appeared to have gone away. Two years later the problems resurfaced and after a training run I returned home where without warning I felt extremely unwell, went into ventricular tachycardia (VT) and was rushed to hospital in an ambulance. I was slipping in and out of consciousness and was later told that my blood pressure was 70/30 with a pulse of 29 beats per minute. My normal heart rhythm was eventually restored and in the ensuing months I had various tests culminating in a catheter ablation, which temporarily resolved the problem. I continued my duties at work and resumed running, albeit recreationally, but eventually the problems resurfaced. Without warning my heart would suddenly go into VT during even the mildest exercise such as walking up an incline and on three occasions when it failed to return to normal rhythm I had to have Cardioversion. Clearly things couldn't continue and I was fitted with a defib and prescribed amiodarone and betablockers.

After recuperation I resumed my police duties, although by this time I worked in a non-front line role. My sporting days were well and truly over, and I "plodded" along until

my retirement from the force in 2009. Shortly afterwards I started to work for myself as a gardener as this activity had presented me with no problems since having the defib fitted. In 2010 it was decided that I would cease taking amiodarone as my device had never "shocked" and my ruddy complexion was now clearly demonstrating one of the side effects of this potent drug. Eighteen months later, whilst out gardening I suddenly felt my heart starting to beat rapidly and when the defib didn't "kick-in" I promptly returned home and rang Jeanette. An ICD download revealed that my heart was in atrial fibrillation (upper chamber problem this time) and I was quickly put back onto amiodarone and given an appointment at the warfarin clinic. Five days later my heart rhythm returned to normal but just a few hours later I had a transient stroke, where I temporarily lost my speech and I was rushed to hospital where I was promptly started on warfarin.

Since then I have returned to my gardening but had to reduce my workload, as I no longer feel capable of prolonged mild exertion. My family and the caring staff at the hospitals have been towers of support to me and I cannot thank them enough. What I have found throughout these experiences is that my confidence was severely dented at each setback, and I think this is a common theme when I talk to people at the group meetings. However I remain positive and thank my lucky stars that I am still here.

Ian Woodward

### DO YOU WANT TO SHARE YOUR STORY?

If you want to share just send your story to George S Davies (georgedavies48@sky.com) or post to 103 Redearth Road, Darwen, Lancashire BB3 2AR



## Sue's Story (in a nutshell)

My heart problems began when I was 19, with a cardiac arrest. This was a reality check for me and my family. Now at 46, I'm still around and kicking and that's in no small part to the wonderful Dr Bennett, Jeanette and team who I can never thank enough.

My first ICD was fitted in 1999. I took this in my stride, until a few weeks later when I had my first shock. While browsing in HMV, I started to feel light headed, then, wham, I heard and felt a huge bang, which threw me back against the CD racks. Confused and panicky, I looked around for a clue as to what had happened. This was Manchester City centre after all and stranger things have happened before. All around me was as it should be, however, everyone was looking at me with varying expressions, as apparently, I had screamed out a rather choice expletive as the air was punched out of my lungs when the ICD did its thing. It actually surprised me as much as my onlookers.

A week later, I received two more shocks while at the tram stop, where I entertained a train full of people and frightened small children at the same time. Of course being a woman I can multi-task. These shocks led to my next surprise. I began to find it difficult in coping with the physical reality of having an ICD and also the psychological aftermath of the shocks. It's not something that people talk about and to be honest, I felt a little ashamed of my reaction. I was very independent before my implant and I thought I was stronger than that. I now know that's all rubbish; it can happen to anybody, even the most confident of person. I started to become someone who was afraid to go out alone and I

began to suffer from panic attacks at the silliest things. However, with a combination of CBT (Cognitive Behavioural Therapy) which helped change my thoughts and behaviours, time and Wythenshawe's Cardiac Rehab relaxation techniques – things did get better. Before I knew it, I was back to my 'normal self'. I'm more than grateful for my life saving ICD and all the help and understanding I received from friends and family. The point I'm trying to make (quite badly) is that, if anyone else finds themselves in the same situation there is a light at the end of the tunnel.

Susan Cunningham

At the meeting with Fabrice on July 31st, Medtronic will be sponsoring a Buffet Lunch which will be served at 1:00pm. So that we know that you will be coming to the Meeting and so that food can be ordered accordingly, **please fill in this slip and return to:**

Sister Jeanette Hornsey c/o Cardiological Secretarial Office, Wythenshawe Hospital M23 9LT

Or email [jeanette.hornsey@uhsm.nhs.uk](mailto:jeanette.hornsey@uhsm.nhs.uk)

Name/s.....

I/We will be eating Lunch.....

### What happened?



On the 13<sup>th</sup> March 2012, during a football match between Bolton Wanderers and Tottenham Hotspur, Fabrice Muamba suffered a cardiac arrest, which stopped his heart for 78 minutes.

### The fight for life



Doctors and paramedics at the scene fought to keep Fabrice alive and in the process administered 15 cardioversions followed by weeks in Intensive Care at the London Chest Hospital.

### Living life



**Come and listen to Fabrice and share his experiences living his life with a critical heart condition and an ICD.**



FABRICE MUAMBA

## WYTHENSHAWE ICD SUPPORT GROUP MEETING

WEDNESDAY JULY 31<sup>ST</sup>

2:00PM

EDUCATION & RESEARCH CENTRE

WYTHENSHAWE HOSPITAL

(Refreshments served at 1:00pm)

# Gene Therapy moves forward



## Support Group Nominated

The ICD Support Group was recently nominated for an Award at the Annual UHSM Awards, alongside two other Groups who do fantastic voluntary work at the Hospital, one supporting patients who have difficulty in feeding themselves and one who assist patients, carers and visitors when they first enter the Hospital. The Award was won deservedly by the Group that supported patients at meal times and many congratulations to them.

## JEANETTE'S CORNER

Hi everyone and welcome to all new ICD patients. I hope you are going to enjoy the Newsletter as much as we all do and please feel free to write a little something to help George continue to put together interesting articles for us to share.

This is a very exciting Newsletter and an even more exciting meeting ahead, with our very special guest speaker Fabrice Muamba kindly agreeing to come along to share his experiences with you all. This great coup is all down to Lindsay's enthusiasm and perseverance! George has requested that you do let me know if you can attend. I am reinforcing this PLEASE, I do need numbers as Medtronic have kindly offered to sponsor a buffet lunch. Please email, telephone or fill out the slip in the newsletter and post it to me with how many of your family, friend etc. will be attending.

Recently I have had a number of queries regarding the DVLA on driving implications when you have an ICD fitted. Normally you will see Lindsay or myself prior to implant, we would always inform you on how long you should not drive for after your device has been implanted.

## IN GENERAL

If you have had a serious heart rhythm/collapse which has caused you to be admitted into hospital, then normally you would be advised to contact the DVLA, as a 6 month suspension is highly likely. If however we discuss your ICD being offer as a precautionary device, i.e. you have not had a dangerous heart rhythm or collapse, then you DO NOT NEED to inform the DVLA but, you should not drive for one month following the ICD being fitted. There are a few of you, who in good faith will have informed the DVLA even when your device was implanted just as a precautionary measure, but sadly this has led to long delays in you being given permission to drive.

If you have any doubts at all after discharge from hospital, do telephone Lindsay or myself for advice.

Have a lovely summer!

Jeanette

## Overcoming adversity



Well, who'd have thought that I'd have made it to forty years old! I've dodged death a couple of times and still manage to come out smiling.

When I was 11 years old, I had cancer in my leg; everything went well I managed to keep all my limbs with a new pioneering technique. The hospital treated me with chemotherapy, massive doses for days on end, every week for a whole 12 months. My life managed to run its natural course, I went to school, college, found a job, bought a house, got married, got divorced!

Then another problem, I had that horrible feeling my head was going to explode. Not all the time, just every time I exerted myself, even walking upstairs to go to the loo was a trauma. So much so, that I invited my Mum to stay with me for a couple of days. One evening I climbed into bed with her saying that I felt like I was drowning when I lay down and I was scared. If you knew me, you'd laugh, I'm not the smallest of people, I'm really outgoing, gregarious and outspoken, but this wasn't nice, or funny. We phoned the out of hours doctor and he admitted me to hospital; it turned out that I had heart failure.

They're the most scary words that I think you can ever hear. I mean, how can you live if your heart fails, surely you can't be alive?? I spent a couple of weeks in my local hospital, yo-yoing through every emotion you could imagine, one doctor said I had to take water tablets, a cardiac nurse told me I'd have to sell my beautiful house and buy a bungalow and get some help in, because when I had my heart transplant I'd need help!!

I was even more confused now, so many different scenarios. At last I was told that I was going to Wythenshawe Hospital and I couldn't wait, at that time anywhere seemed better than where I was. That's where I met my guardian Angel, with her silver hair, blue smiling eyes and loving warm arms when you needed that hug. Jeanette, I couldn't imagine a lovelier person to talk me through the procedures of then and what was to come.

Well as you can imagine, I'm okay now life is great! On a recent holiday to Mexico, I swam with Stingrays, Sharks and Dolphins; I flew in a helicopter – which was the fastest and most exciting 15 minutes of my life. I go to concerts with my friends, enjoy nights out, work full time and have a beautiful dog called Chester who keeps me very busy with walks, playing tug and just snuggling on the sofa. Oh yes, and I love driving my Mini Cooper – absolutely nothing keeps me in, well possibly 10" of snow and the thought of clearing my drive with my snow shovel again!

I suppose, this story is all about living and having something to live for. There are so many things out there I've not done or seen yet; I haven't been for a ride in a hot air balloon (it's booked for April), seen the Sydney Harbour Bridge, been on safari in South Africa. There are so many things that I've done, but too many that I have yet to do.

Possibly the most wonderful even this year will be my wedding to Phil in September and an amazing honeymoon in Las Vegas to follow . . . dreams really do come true and you should never give up on your dreams

Alison Clayton

Two clinical trials are planned for a few dozen British patients who will be deliberately exposed to a virus carrying a synthetic copy of a human gene known to be involved in boosting heartbeat.

The first trial will be carried out at the Royal Brompton Hospital in London and the Golden Jubilee National Hospital in Glasgow. The patients will be part of a group of 200 from around the world who will have the virus injected via a cardiac catheter inserted through a vein in the leg. A second trial at the Harefield and Papworth hospitals will be based entirely within the UK and involve 24 patients with chronic heart failure who are already fitted with an "artificial heart" known as a left ventricular assist device, which helps to pump blood around the body.

The aim in both trials is to inject additional copies of a healthy gene, known to be responsible for a key protein involved in regulating the rhythmic contraction of the heart muscle. It is hoped that the extra genes will remain active within a patient's heart for many months or even years.

Scientists believe the approach could lead to a significant improvement in the efficiency of the diseased heart to pump blood around the body – so improving the quality of life of thousands of patients with progressive heart failure who develop serious ailments as well as severe fatigue.

Scientists warned that it will still be several years before the technique can be made widely available. They do not want to raise hopes unduly as many previous gene therapy trials on

patients with a range of other illnesses have failed to live up to expectations.

However, the heart researchers said they are optimistic that the gene technique will improve the quality of life in at least some of the patients, who would otherwise suffer deteriorating health and life expectancy – a third of patients die within a year of diagnosis.

"Once heart failure starts, it progresses into a vicious cycle where the pumping becomes weaker and weaker, as each heart cell simply cannot respond to the increased demand," said Alexander Lyon, a consultant cardiologist at the Royal Brompton.

"Our goal is to fight back against heart failure by targeting and reversing some of the critical molecular changes arising in the heart when it fails."

The gene-therapy trial involves the injection of a harmless virus, called adeno-associated virus, which has been genetically modified with the SERCA2a gene responsible for a protein that stimulates contraction of heart-muscle cells.

Dr Lyon said extensive safety work has already been conducted on the virus and it has not been found to cause health problems in other gene therapy patients.

Once the virus has delivered the SERCA2a into the muscle cells, it is quickly degraded, he said.

Professor Sian Harding of Imperial College London said that extensive research on the SERCA2a gene has shown that it can be safely inserted into heart-muscle cells with the effect of boosting the size and speed of contraction. "It's been a painstaking, 20-year process to find the right gene and make a treatment that works, but we're thrilled to be working with cardiologists to set up human trials that could help people living with heart failure," Professor Harding said.

Professor Peter Weissberg, medical director of the British Heart Foundation, said gene therapy shows great promise in providing a new way to treat heart failure, but warned that there is still a long way to go.

"There was a terrific fanfare around gene therapy about 10 to 15 years ago. It was going to cure everything and, like all things in science, it wasn't that simple," he said.

"This project is a great example of the slow burn of good laboratory science translating into potential clinical treatment. We don't know if it's going to work yet, we all hope that it will."

*Reprinted verbatim from The Independent Newspaper on April 30th 2013.*

*The article can be located at: <http://www.independent.co.uk/news/science/gene-therapy-to-offer-up-to-1m-heart-patients-new-lease-of-life-8595621.html>*