

# Insulinoma

*Personal stories from real people*



**Insulinoma:**

Is a rare tumour derived from the beta cells of the pancreas producing large amounts of insulin

## My Insulinoma: Personal experiences from real people with an insulinoma

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This Insulinoma eBook is dedicated to the memory of Te Rina Lytle who died at the age of 15 years old from a malignant insulinoma

## Preface

Insulinomas are a rare type of functional neuroendocrine tumour usually found in the pancreas. They are called functional because they produce excess insulin which can cause hypoglycaemic symptoms and ‘episodes’.

Hypoglycaemia is a condition where blood sugar levels in the body are too low and a variety of symptoms can occur. A hypoglycaemic episode can cause many symptoms including anxiety, sweating, hunger, headache and tingling or numbness of the skin. If low blood sugar levels are not treated promptly, hypoglycaemia can lead to coma and death.

Even the smallest tumours can cause symptoms connected to hypoglycaemia insulinomas are rare and for many it can take a long time to get it diagnosed. This book has been created to provide a greater understanding of insulinomas and contains information regarding symptoms, diagnosis and finally treatment of this rare condition. This book is written by those with an insulinoma who want to share their story about having the condition. It is written in their own words and in their own voice in a series of personal stories and experiences

The intended audience for this book is professionals in the medical field who are involved in diagnosing and treating insulinomas. It is also for those who suspect or are diagnosed with an insulinoma and require further information or an understanding of this condition. This book can also be useful for people who have to care for and live with someone who has an insulinoma.

A special thank-you goes out to all those who have been on this journey and who have shared their experience of what it is like to have an insulinoma.

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## Part 1

### Our stories

#### **Fleur Horslen, Essex, UK – Benign Insulinoma, 2012**

*“For thirteen years I’d been told that my symptoms didn’t fit any illness...”*

My story starts in the summer of 1995. Before that I had enjoyed robust health, was a size 10 and weighed 9 stone. The first “funny turn” happened when I was at home and my brother was downstairs. I became confused and couldn’t understand the noises he made and what he was doing. I had a few more attacks and thought I was going mad. Finally I had one of these “funny turns” at work and my manager sent me to the doctor. The doctor said it was probably just an infection and gave me antibiotics. However the attacks still happened and I saw the doctor again. Blood tests showed that my blood sugar was low and I was having hypoglycaemic attacks (hypos). I was advised to eat half a chocolate bar when I was having a hypo.

After a year or so of these turns I found that I could control them through diet. The main triggers for my hypos were too much alcohol or missing a meal. For a long time I believed that it was my fault, that I was doing something to create these hypos. The hypos became more frequent and I became embarrassed about having them. I wouldn’t tell anyone and would consume something sugary as quickly as possible so people wouldn’t realise.

I struggled for 13 years to manage my hypos. My treatment for hypos was orange juice and a packet of biscuits or something sweet. When you are having a hypo it is difficult to control how much sugar you eat as your body craves it to rectify the problem. I would eat until I felt better. The result of this “treatment” was a weight gain of 3 stone and a size 14-16 in clothes.

I went to see a private endocrinologist and after several blood tests was told that medical science had failed me and that my low blood sugar wasn’t a problem. I left upset thinking I was making a big thing out of nothing.

In 2008 I developed neurological problems. I was dizzy all the time, unable to speak properly, couldn’t walk in a straight line and my left side was weak. I didn’t think it was blood sugar related as I was not having my normal hypo warning signs. The attacks became worse and I was admitted to A&E. When I told the admitting doctor about my symptoms and my low blood sugar he told me that they would only treat one thing and I had to choose which one! I opted for the neurological symptoms and was referred to a neurologist. The neurologist discharged me with a diagnosis of migraines and stress telling me that I had made up my symptoms. I left hospital wondering what kind of person I was if I could make up my illness yet it seemed so real to me.



From 2008 until 2011 I was permanently unwell with balance issues, a permanent headache, depression and left sided weakness. I put on a further 5 stone meaning I was now obese. In the summer of 2011, I started to have episodes where I would lose my memory and think that I was in a dream. In one of these episodes I passed out and ended up with a black eye. I also had severe migraines and had to call out the emergency doctor. My GP referred me to another neurologist but again despite running several tests he was unable to come up with any other diagnosis than migraines.

In October 2011 my GP took several blood tests, one of which was fasting blood glucose. My blood sugar was 2.9. The test was repeated and this time the result was 2.1. I was then referred to Dr Toh, an endocrinologist at the local hospital.

I will always be grateful to Dr Toh. She was the first person to listen to me and take my blood sugar problems seriously. I took my blood sugar readings which indicated that I was permanently hypo. Dr Toh was certain that it was an insulinoma. As I had experienced blood sugar problems for so many years it was likely that it was benign and in fact this was the case. Tests confirmed that I had a 1cm lesion in the head of my pancreas. Apparently I was a text book case. My husband and I just laughed. For years I'd been told that my symptoms didn't fit any illness and here I was a text book case. I guess we had just been looking in the wrong textbook all these years!

I was referred to Dr Simpson at Addenbrookes Hospital who was brilliant. She wanted to run the tests again to ensure that the diagnosis was correct. I was admitted to hospital as my blood sugars were unstable and that I was now hypo unaware, meaning that my body was so used to hypos that it didn't react when my blood sugar dropped.

The 72-hour fasting test confirmed low blood sugars with abnormally high insulin levels. An endoscopic MRI showed the lesion clearly. Unfortunately, it was in the head of the pancreas so I needed to have a Whipple operation in which the surgeon removes the head of the pancreas, the gall bladder, duodenum and part of the stomach. It was a major operation with many complications but I had no choice. I had been told that if I did not have the operation it was possible that I would have a massive stroke, heart attack, fall in a coma or die in my sleep.

I had my surgery on the 21<sup>st</sup> May 2012. The operation took over 11 hours. When I woke up I had so many drains and wires that I didn't dare move. I had my own nurse to look after me the whole night. Incredibly, my blood sugar levels were normal straight away.

I spent one night in recovery, two nights in ICU and two weeks in a surgical ward. I found the recovery very hard. I was in pain, constantly being sick and very tired. I developed pneumonia and my wound opened. The multidisciplinary team were excellent. On a daily basis I saw the surgical and endocrinologist teams as well as dieticians and physiotherapists.

Being discharged was a major step in my recovery. I made the effort to get dressed every day and found that I could eat and drink without being sick. The hospital had arranged for the district nurse to come in daily to wash and change my wound and I appreciated their support during this time.

The post-op support from Addenbrookes has been fantastic. Dr Simpson and the surgical team will see me on a regular basis to ensure all is well.

I am now 11 weeks post-op. I still take painkillers and get tired very quickly. I have friends doing the ironing and cleaning for me and most of our meals are ready meals. However I love not having to eat all the time and my confidence is starting to grow again. I have also lost four stone without any effort.

As I've had hypos most of my adult life, it is difficult to know what my life will be like without them but I'm looking forward to it!

## **Angela Shaw, Essex, UK - Benign Insulinoma, 2012**

*“I could not work out how to write some words. I tried to use a computer... but could not focus on the screen.”*

In September 2011 I had just moved from Essex to Berkshire to study for a Master’s degree in Conservation and Forest Protection and I was excited about the prospect of change and all the new things I would be learning and experiencing. Unfortunately, the year ahead was not to be as enjoyable as anticipated.

Only three weeks after moving, and at the end of my first week at university, I passed out in the morning whilst out walking my three dogs. I do not remember how we all got back home. I only had a few images about the journey and I initially thought it had been a dream and that I had not even left home. It gradually dawned on me that I had indeed gone out and had probably appeared drunk to bemused passers-by. Falling into hedges and crossing a road without looking or putting dogs onto their leads is not the done thing! Needless to say, I was feeling very tired and confused at this point. My new GP kindly fitted me in that morning and after a thorough check decided that I must have tripped and suffered concussion (I had grazed my face and badly bruised and cut my knee). Subsequent blood tests did not show anything either.

This preceded a month of very strange occurrences. Mornings were particularly difficult. Often even three alarms would not stir me and when I finally got out of bed I could not work out what I should be doing. Sometimes I could not even tie my laces. Symptoms included pins and needles in my mouth and face, sweating and heart palpitations after only slight exertion. Goodness only knows how I got myself to uni every day. However, as the day progressed I started to feel better and was able to carry out tasks much easier in the afternoon / evening. Nonetheless, I started to dread going to bed as I knew how I would feel in the morning. If I engaged in anything more than gentle physical activity I would become, at the least, shaky, emotional and disorientated. I consulted Accident and Emergency, NHS Direct and also another GP but responses ranged from ‘It takes time to recover from concussion’ to ‘There is nothing wrong with you’.

One morning I woke up and did not have the usual fuzzy head. This filled me with optimism as I thought I was finally getting better. Unfortunately, I deteriorated through the morning. I could not focus on the board in lectures and taking notes was tricky, I could not work out how to write some words. I tried to use a computer during break but could not focus on the screen. I went to see the course administrator and was in tears but did not know why. She got me to sit down and went off to get a first aider. Next thing I knew I was sitting in the back of an ambulance. I had passed out again for about five minutes. Fortunately for me, the paramedics had the presence of mind to check my blood glucose (BG) and this started me on my way to a diagnosis.

After a fasting (overnight) blood test returned a 1.8 BG my GP referred me to a brilliant endocrinologist who immediately suspected an insulinoma and got me into hospital for a 72 hour fast, CT scan and MRI. I was not allowed to drive for a couple of months until I had been on

Diazoxide for a while and BG was controlled with this and eating regularly. Over the months there were many visits to hospital for other blood and urine tests and ultimately a referral to Oxford. Despite the initial flurry of activity, it was a very trying and protracted wait. The hospital did not carry out the tests properly and I had to repeat the fast; another laboratory technician took it upon themselves to carry out alternative tests to those requested as they were not 'usual'; I was given an incorrect container for the urine test etc. Somehow, given multiple setbacks, I did not get to Oxford until March. They then wanted to redo all the tests along with a calcium stimulation test and different MRIs: Further delays but they were very thorough and professional. I found this period of uncertainty very difficult even though my symptoms had subsided. Nobody knew why my BG had stabilised and would not fall so quickly if I did not eat. Looking back over the years I think that the illness had been cyclic as, with the benefit of hindsight, I could see periods of unexplained illness. I also put on over three stone in weight. Thankfully, this is now coming off.

I finally had my operation at the end of June 2012. They performed a distal pancreatectomy and removed my gall bladder (as it was full of stones) via open surgery. I had not had any symptoms from the stones but they had been noticed on an MRI. Recovery went well and I went home after a week. I managed to look after myself and got out twice a day for a walk, gradually increasing the distance. I think this really helped the healing process. Pain was not an issue. Of course it was uncomfortable and full movement was impossible but it was bearable and so long as I did not overdo things, it was fine. I had an epidural for the first few days in hospital and I was then on codeine and paracetamol. I was off these by the end of the second week. I did suffer from acid reflux though but this eventually subsided and I controlled it by taking cider vinegar before each meal. Life was pretty much back to normal after 2-3 months although I still get tired and my tummy feels 'compressed' sometimes. I think I will have the numb feeling for a while yet in the area below the incision (which is still slightly swollen subcutaneously).

I very nearly gave up my course in December as I was so tired and found it difficult to cope with the demands of study. However, I decided to stick with it on the condition that I lowered my usually too high expectations of myself. I am glad I did this as it gave me something else, other than the illness, to focus on. If you are going through the process of diagnosis, try and stay positive, there is light at the end of the tunnel; it does get better and you will get your life back. I tried to eat as well as I could (vegan) and looked after myself with complementary therapies. It is all about what works for you as we are all so different. Wishing you well on your journey.

## **Kori Woodard, Banff, Canada - Benign Insulinoma, 2011**

*“I would hope that by telling these stories, diagnosis and treatment can be faster”*

Funny how you can't forget some dates, no matter how hard you try. I would count the day I had insulinoma surgery as one of these, May 17, 2011. Likely difficult to forget as it had been a long time coming and life altering.

Looking back on it, the first time I remember some symptoms was when I was pregnant with my second child in 2007. I was able to stay active throughout my pregnancies but was feeling particularly tired with my second. I was out cross-country skiing one day and had an overwhelming feeling that I could not make it back to the trailhead. Eating anything seemed out of the question as I thought it would make me more tired. I did make it back and chalked the fatigue up to pregnancy. My glucose tolerance test during my pregnancy was at 3.2.

After the birth of my second child, I went on to do a half-marathon when he was 4 months old. Again, I felt over-the-top fatigue, headaches, and body ache for a long time after running. At the time I didn't recognize these symptoms as too much of a concern. However, as time went on I found I could do less and less without feeling fatigued. Christmas day landed me in the emergency department with a mystery illness after skiing, characterized by flu like symptoms, painful legs, cold sweats, and shakiness. I was treated for suspected mastitis and put on antibiotics.

My reaction to the fatigue was to do more exercise and live “healthier” through better nutrition. By late 2008 and early 2009 I definitely knew something in my system was off kilter. Chronic fatigue syndrome was suggested as well as hypothyroidism (runs in my family). I was put on synthroid to see if it helped. I completed two mini-triathlons but was slow and wiped out by both. By this point I was getting a bit desperate as it seemed the medical system had little to offer, so I sought out alternatives to the conventional medical system. Thus began my visits to a psychologist (to see if I was depressed), a homeopath, and a medical practitioner who practiced holistic medicine.

My frustration grew as no-one seemed to be able to help. I continued seeing my family doctor occasionally but she didn't seem to have much to offer either. The alternative system suggested I needed “energy balancing” and that my hormones were not balanced (close to the truth but they focused on estrogen and testosterone). I was finally sent for a brain MRI in 2009 as I was having tingling in my extremities and odd neurological symptoms. The MRI did not show too many abnormalities.

During 2009, I started have episodes during exercise where I would be unable to stand up and my legs and arms would shake. These appeared to my husband to be some type of seizure. I was usually tired and emotional afterwards and sometimes did not remember as much time as he did. I stopped making plans to do any major activities or exertions and even felt too wiped out to

plan family holidays. The relationship of these 'episodes' to food was unclear, often I would feel the worst a couple of hours after eating or if I was trying to exercise.

During 2010, I was referred to a number of neurology clinics. Every practitioner that I saw had a waiting period to get into see them, and I would tell myself that it would be figured out at the next appointment, but never got any answers. I did visit the emergency department a couple more times but they didn't take a blood sugar. Sometimes the 'shakes' would happen when I took my kids swimming, going for a walk with my family or after work. Whenever I tried to ski I would usually pass out in bed afterwards with the cold shakes and 'lose' a couple of hours while I slept. Eventually I stopped doing so much and was even tired with daily activities. Teaching courses at work was difficult as I sometimes felt as if I didn't make any sense or was slurring my words.

I had switched family doctors and my new doctor seemed willing to pursue other hypotheses for the symptoms I was having. I had a second MRI on my brain which showed no change. I was fed up with the system at this point and even avoided making appointments. I had given up in some ways and was very frustrated and tired. In February 2011 I had an absence seizure that I wasn't even aware of. My family happened to be there and informed me I had been talking to them and shaking for a period of time. At the urging of my husband I saw my family doctor. She strongly felt it was epilepsy and referred me to an epilepsy clinic. At the same time I asked her if it could have anything to do with blood sugar. She agreed to test my fasting glucose one more time. A few weeks later I went in to get the test, even though I felt normal (although a little tired), my blood sugar was at 1.9. My doctor immediately suspected insulinoma and sent me to an endocrinologist. Even though the 'cure' involved major surgery, I was so relieved to hear an explanation for all my symptoms. An MRI scan with contrast dye (Gallium maybe) confirmed that the insulinoma was between the body and the tail of the pancreas near the pancreatic duct. Coincidentally (or not), a neighbour in my small town of 8,000 had also been through an insulinoma operation in 2009.

While waiting for surgery my blood sugar continued to drop dramatically, as low as 1.6 occasionally. On a subsequent visit to emergency they put me on Diazoxide, I found it raised blood sugar in general but with any exertion, sugars would fall just as dramatically as before. It didn't seem to fix the fatigue at all.

The operation took 5.5 hours and was attempted laparoscopically but because of the location of the insulinoma my surgeon also opened a hand port. I felt so lucky to have a wonderful surgeon and be able to regain my health and my hope after the surgery. The surgery has made a huge difference in feeling clear headed, being active and being present for my family. I empathize with anyone still struggling to get diagnosed or with the complications an insulinoma can bring. I would hope that by telling these stories, diagnosis and treatment can be faster.

## **Sally Wheeler, Newcastle, UK - Malignant Insulinoma, 2011**

*“... The oddest period of my life”*

In June 2011 I had a private gastric bypass, the best money I've ever spent. I saved very hard, had it done, lost the weight and felt fabulous. But in about the October 2011 I started to have what I can only describe as white splashes in front of my eyes, just like a drop of water but white. When it affected my reading ability I went to the opticians, but they found nothing untoward.

I muddled through for about two months then had an appointment to see my bypass surgeon. In the meantime, I was getting tingly lips, hands, even around my midriff and down my arms. I explained this to my surgeon and he thought I may have Nesioblastosis, which is a rare side effect from bariatric surgery. 'Great' I thought, I pay all this money and get this really horrible illness; I had to go through some tests to establish if it was that or not.

A month went by, by then I was having regular hypos; feeling tingly, confused, I couldn't see properly, I was acting strangely and became very vacant. I couldn't walk far or straight without stumbling. Once, I nearly knocked this poor woman flying in M&S, I think she thought I was drunk! My poor husband, David was really worried as I was going into low sugar mini comas. We didn't know at the time what they were until I could not be woken one Sunday morning: I was having a fit, my eyes were staring, and I was shaking but couldn't be woken. David then rang 999. They sent out paramedics who gave me a glucose drip. Within 10 minutes I was lucid but didn't know why or what had happened.

It happened again three more times, within three weeks, always on Sundays or Mondays which were my days off work. I was always so tired that I would lie in on my days off which in turn dropped my sugar level as I hadn't eaten for at least 12 hours. My sugar would go down to about 0.5. David rang our wonderful GP to tell him what had happened and how often. My GP then sent me straight to an endocrinologist, Dr Bennett, who did the 72 hour fast. The fast only lasted about 20 hours. On the first night, I had my blood tested every hour; it was low all night so they put me on a glucose drip. When the specialist nurses Jackie and Phil came in the next morning the drip was removed and they waited an hour or so then took my blood every 15 minutes for about one hour. They said they had never seen sugar drop so quickly and so low. They felt it was dangerous to go on any longer as they now had what they needed, so I was fed and watered. I rang Dave then went home. Within a week I was on Diazoxide, had all the scans done and by March all the results were in. I had had a couple of more blood tests by then also (I had that many can't remember the names of them). I'd seen Mr Senn and was told I had an insulinoma which were nearly always benign. I had never heard of it and since the Diazoxide was working could I just stay on that and get on with life? "No" he said, it had to be removed as it was a functioning insulinoma and I would need open surgery as I could lose my spleen if I had key hole. I said I didn't want it done until July as my daughter was getting married in May. I needed

time to think also, I didn't want it done and was being a coward; "OK" they said and we settled on a date in July but by the beginning of June they wanted to do it sooner, so June it was. I had a distal pancreatectomy and all went well; I spent two weeks in hospital, went home, then got my histology results. The insulinoma had been cancerous and it had gone to my lymph nodes which they had removed. I was very shocked but they assured me it had less than 1% chance of coming back, so four months down the line I'm getting ready to go back to work and I feel positive as I have no more horrible hypos. My energy levels are coming back and I now feel happy and optimistic. I hope this has helped xx



## **Sarah Vivian, Cork - Benign Insulinoma, 2010**

*"I would go off into fairy land and be 'absent' for minutes at a time"*

I first went to the doctor with eye problems: difficulty focusing and double vision, and I found when sitting in car as a passenger that I felt as though the car beside me was moving, when it wasn't. I fell a lot - my legs were giving way. I had put on a fair bit of weight and couldn't shift it, I called into a friend on my way back from Weight Watchers and had a cup of coffee and went very hyperactive - rabbiting on and making little sense. Then I had a seizure at night, my husband called an ambulance but by the time they arrived I was fine. A few months later I woke up one morning, got out of bed and fell over; I realised I couldn't stand up and couldn't move any of my left hand side. When I spoke to my husband to say what had happened I could only slur and my face was also paralysed down one side. Off to casualty, where I was tested for everything - except blood sugar! About four hours later a nurse offered me a cup of tea and a sandwich and I miraculously recovered. My GP looked back at my notes and referred me to a neurologist - a suspected brain tumour. After being wired up and my brain tested - it showed abnormalities on the left hand side but nothing really conclusive - a brain scan was clear. She suggested it might be an abnormal sort of epilepsy and put me on pills. After two weeks you have to have thorough blood tests to make sure you aren't reacting to the medication - my blood sugar was 2.3.

I was referred for a glucose tolerance test - the nurses kept popping out to look at me and asking me if I was feeling ok - "Fine" I said, bemused. They had never seen such an extreme reaction. My GP said it was a possible insulinoma and referred me to the local endocrinologist for my first 72 hour fast.

Not fun - I came to whilst wandering down a corridor - no idea where I was or how I had got there - I was offered a cup of tea and went very silly, giggly and felt very uncontrolled - the nurses and a doctor were laughing at me and my antics. No one took any blood. My GP came in in the middle of all this and also laughed at me. After the 72 hours the endo said to eat a sandwich and I would be fine and yes I could drive. My husband, luckily, came to collect me and I was a total mess for about a month - the worst I had been, unable to drive or walk without keeling over. And the endo said he wasn't sure what it was but didn't think an insulinoma. When I felt a bit stronger I went back to the GP and got very cross. I said it was a disgrace that I was laughed at, especially by him as he knew me well and knew that that was not normal behaviour for me. I insisted on being referred to a top endo in Sydney and I had made enquiries and I gave him two names to check out for me. I was referred to Dr. Clifton Bligh in the Royal North Shore.

What a difference - a lovely man who was very thorough - MRI, CAT scan, 72 hour fast and angiogram with calcium stimulation test. This was in 1993 and it was a very new test then - apparently I was the second person in Australia to have it. He told me that it was almost certainly an insulinoma, possibly several, but the pancreas was a tricky place to get to and nothing was showing on the MRI - only the calcium stimulation and the blood test so he wanted to wait and

test me again at intervals. He sent me to a dietician who advised me on what to eat and how often.

My life was transformed - no more hypos, I even lost a bit of weight, I started to dream again (I very rarely dreamed). I went for check ups every six months and he was pleased with how I was coping so decided to keep waiting.

In 1997 we moved from Australia to Ireland and I took all my medical records with me. With the stress of moving I had put on weight again and had more hypos. New endo read my notes and said he wanted to repeat the 72 hour fast. Said he did not believe I had an insulinoma as they were so rare. Did not tell me what I had. He told me to lose weight and I would probably find my symptoms disappeared again. Like a mad woman I went back to him every 6 months for the next 3 years and had another 72 hour fast. I also asked for, and he did actually agree, to another MRI scan. Nothing.

Now ten years since my first diagnosis and I had been told that it was unlikely I would conceive again due to my low blood sugar and as I was now 40 I got slack about contraception - I was pregnant within two months! The first month was terrible - I was very worried and had a lot of hypos but after that I felt wonderful - really strong for the first time in years - I only put on about half a stone and had a trouble free pregnancy. My blood sugar was 3.2 to 3.8 and to be fair they did check it regularly. After my daughter was born in 2003 (the boys were now 11, 12 and 14) I was very bad - very unstable and we also moved house when she was five months old.

In the middle of moving I had a very bad seizure and my husband couldn't wake me - he called the out of hours doctor who administered a glucose injection and I came to. No one took my blood sugar as the doctor couldn't find her machine and felt it was more important to jab me quickly and mine was in the other house. Back to the endo - he had retired and a new one was equally dismissive - "Oh I expect we all have blood sugars of 3.1 this morning with the mad house this is" she said. She wanted to do another 72 hour fast - I refused.

No doctors for five years, I muddled through. Then my closest friend died very suddenly in early 2010 of a heart problem. She had had this for years and was due to have an operation but died too soon. She had a seven year old son at school with my daughter. After losing her I realised I had to get this sorted - she nagged me constantly in my head for the next few months and, after I was made redundant in June 2010, I asked my GP to refer me to another doctor - a physician in Bantry who I knew was brilliant but who was not a specialist endocrinologist. He read through my notes, said it was ridiculous I had had to live like this for so long, told me NOT to attempt lose any weight and referred me straight to the surgeon in the Mercy Hospital Cork without any further tests. A month later I was in Dublin having an endoscopic ultrasound where the insulinoma was located and in November (the day after my 49th birthday) I had the operation - a Whipple. Brilliant surgeon, brilliant team, lovely nurses. All so kind and thorough.

Thinking back on it I had my first symptoms after my first child was born - in 1988. I had another two children in quick succession and the symptoms only really got bad when I was really trying to lose weight after the third and I wasn't being entirely sensible about it - missing meals and over exercising.

My symptoms started with unexplained tripping over things, behavioural changes (after caffeine), feeling drunk and over excited and giggly after very little alcohol (I thought I must have had more than I was sure I had had), vision problems - like looking at a bright light, and double vision and vertigo. Then I started to get vague and would lose track of conversations, I would find that I couldn't make sense and found it hard to articulate what I meant sometimes. The falling over got worse. I would go off into fairy land and be 'absent' for minutes at a time. My face would twitch and mouth tingle. I very rarely got hungry but would crave salt (perhaps an adrenal thing). The salt cravings have gone now. I just felt very weak and feeble and always very easily exhausted.

The diet, as given to me by the dietician in the Royal North Shore Hospital, Sydney, was small meals every 2-3 hours (two hours when trying to get stable but never more than three and half hours). Low GI (glycaemic index) - so brown rice, pasta, brown bread (I found rye bread the best), oats, all veg are ok but go easy on potatoes and sweet potatoes. Most fruit is ok but try to steer clear of bananas, melons and grapes as they can be too sugary. Meat, fish are ok. Dried fruit can be too sweet, nuts are good. I was told that if you must have something sweet, have vanilla ice cream, the fat in it helps to slow down the sugar, it has protein in the milk products which is good for also slowing down sugar release. Avoid sugar, caffeine, excess alcohol. I discovered for myself that vinegar was terrible - I read on the internet (yes, I know it isn't always accurate) that vinegar had been used to lower blood sugar in diabetics and it had worked (but tasted disgusting) so I tried avoiding it and improved hugely.

I did manage to live with it fairly well due to the excellent advice of the Sydney doctor and the dietician and the support of my husband and children who were brilliant with me and recognised the symptoms and would be ready with cheese and biscuits or milk. If I was very strict with what I ate and when, I could cope - but of course I wasn't always very good and I paid for it then.

Post Whipple - strong, clear brain, clear skin, dreaming again, so much energy, new business, lost five stone. No point in regrets - I am just living now and enjoying it.

## **Laura Jones, Worcestershire, UK**

**Diagnosed with two insulinomas in January and March 2010, one further insulinoma July 2011**

*“I knew I’d had a funny turn but had no idea about the scale of what had happened”*

My symptoms first started back in September 2008 (I was 25 at the time). It was whilst spending a day in London I noticed that I was feeling drunk. I knew I was only drinking lemonade but still asked my dad if he’d mixed in some gin, which of course he hadn’t! We went for a meal shortly after that and I soon felt a lot better.

My symptoms became more and more apparent and by May 2009 I was experiencing the following regularly; dizziness, lack of concentration, exhaustion, tingling mouth and lips (particularly after eating), severe memory loss and the need to eat regularly as I was constantly hungry, which resulted in weight gain. It got to the point where I wasn’t waking up in the morning, my husband would often attempt to wake me but, despite my eyes being open, I gave no response. He realised that I needed something sweet to eat to bring me round gradually. I struggled with even basic day to day tasks as I felt so exhausted. Exercise was becoming increasingly difficult as I often experienced blurred vision and would feel very unstable on my feet.

I was regularly visiting my doctor to provide updates, in particular that I was having strange attacks whereby I would experience erratic involuntary limb movements (both arms and legs) which appeared to look like a seizure. This was starting to happen in public places, on one particular occasion when I happened to be in a coffee shop during peak time but thankfully my husband was with me and I didn’t injure myself. In May 2009, given the similarity of the symptoms, my doctor advised me there was a possibility that I could have epilepsy and I was referred to a neurologist as soon as possible to look into this further.

On 2nd June 2009 I was following my boss home after work, as I was going to see his new baby boy. On route I remember following him and then all of a sudden my foot went down on the brake, I was trying to control my driving but it was almost like I couldn’t control my feet. My boss in front noticed and therefore slowed down, I managed to catch up with him and he turned right. From here on in, the rest is a complete blur, I don’t have any recollection. The next thing I knew I was parked on a grass verge with my boss stood next to the car looking very worried. I had my head down on the passenger seat and I burst into tears. I knew I’d had a funny turn but had no idea about the scale of what had happened. We then left my car on the grass verge and my boss drove me home. My husband opened the front door and I was very upset as I was unsure what had happened. Thankfully, just that day, I had told my boss about the possibility of having epilepsy so he was aware of what I was going through (to a certain extent). He told my husband in private that I had actually driven into the back of his car and he could see that I was unable to

control my vehicle. I was in total shock when he told me; I felt like I was losing control but couldn't understand why.

I saw the neurologist a few days later and I was sent for an MRI scan and an EEG. I returned to get the results of the tests but they didn't show anything abnormal, apparently this can be quite normal for people with epilepsy as they often needed to catch a seizure during or shortly after it had taken place. Our wedding was pending so it was decided that I'd start on some epilepsy medication and monitor how I coped, increasing it weekly until I reached the correct dose. At this stage I voluntarily surrendered my driving licence. I didn't notice many changes from the medication although I was becoming increasingly tired, but just put this down to the increased medication I was taking. During this meeting the neurologist did recommend that I also saw an endocrinologist just to rule anything out from their point of view, which was sparked following the confession of my constant hunger and the need to eat regularly.

One of the most frightening occasions was in August 2009 when my husband and I arrived at my mum's house; I knew the code for the alarm so she told us to let ourselves in. It took me a while to even get the key in the door and I felt something was wrong. As soon as we got into the house the alarm was going off and I looked at the key pad to enter the alarm code and it was as if I had never seen it before, I had no idea what the number was. I just stood there in a daze whilst my husband tried to engage me in conversation. He called my mum to get the code and I went to sit down. I was feeling so tired so I went to lie down. In hindsight this was the worst thing I could have done as I needed to eat to bring my blood sugar back up. I went into a deep sleep and two hours later my mum and husband woke me up for tea. I was apparently very vague and had a blank expression. I then had two hypos which lasted about 20 minutes each. This was the first time my mum had seen it so it was obviously distressing. When the first hypo lasted longer than usual they called an ambulance, as the ambulance staff arrived I had the second hypo so they were able to witness. They said that this wasn't characteristic of an epileptic attack and they checked my blood sugar level which was 1.9. I was given a sugary cup of tea and some bread coated in jam but, as a precaution, given how low my blood sugar was, combined with my dazed state of mind, I was taken into hospital and kept overnight for further investigations.

I visited my doctor again at the start of September 2009 after my time in hospital and it was during this appointment that the first discussion of insulinoma was raised. My doctor mentioned that whilst she was at medical school she had learnt about insulinomas and the symptoms. The doctor said that she would notify the neurologist about her thoughts and take it from there.

I then got married and we had a wonderful day and a wonderful honeymoon! Upon our return my symptoms got increasingly worse and I was now seeing an endocrinologist. I had a series of fasting blood tests and the lowest it went was 2.6. I was now adamant that I had an insulinoma and so continued on my quest to get a diagnosis. The endocrinologist was very helpful and now that he knew that I didn't have any additional stress I had a prolonged fast which lasted about 6 hours, by which time my blood sugar had dropped to 2.4 and after analysis it showed increased

C-peptide and insulin levels, by this point an insulinoma was looking increasingly likely. In early January 2010 I had a CT scan which confirmed a 2.6cm lesion on the tail of my pancreas which was confirmed as an insulinoma. I was shocked but also relieved at the diagnosis and thankful that after the journey I had been on I had reached a point whereby it could be dealt with. I was put on Diazoxide in the short term and I then had an endoscopic ultrasound to confirm the location.

During March 2010 I had open surgery to remove the insulinoma (at this point the original site of the insulinoma was ruled out and an insulinoma at the head/neck area of the pancreas was confirmed). Although this was successful, the original site of the insulinoma, on the tail of my pancreas, was in fact another insulinoma and so my symptoms remained. This was then successfully removed at the end of April 2010 as well as my spleen and although I had had two lots of open surgery in close succession, I was relieved to see that my blood sugar levels were slowly returning to a normal level despite a few peaks in my sugar levels as my body readjusted. I had some complications following both lots of surgery which included infections, a collection of fluid where my spleen used to be and a build up of fluid on my left lung. All of these were treated following a few returns to hospital.

I continued to have regular check ups and made slow but steady progress with my recovery. During one of my visits to the endocrinologist the genetic condition MEN1; Multiple Endocrine Neoplasia Type 1 was discussed (please see [www.amend.org.uk](http://www.amend.org.uk) for full details of the condition) and it was suggested that I should be tested as I had had multiple insulinomas. Thankfully, I had read an article on MEN1 so had some background knowledge. I agreed that this was worth investigating and an appointment was booked to see the genetics team. It began with genetic counselling which explained all about the condition and my family tree was reviewed to see if there had been any particular health issues that could be connected to MEN1. I had the genetic blood test and had to wait a number of weeks for the results. The results arrived through the post on the morning of New Year's Eve 2010. It was confirmed that I had the genetic condition MEN1 and there was a booklet enclosed explaining the condition. To say I was shocked was an understatement and it really did take some time for the news to sink in. No longer was I dealing with two insulinomas that had been removed, I now had a lifelong condition to consider. Since then, both my dad and uncle have been diagnosed (my brother was clear).

During 2011 I was found to have one further small insulinoma on the pancreas which was picked up by a scheduled endoscopy (I also had another suspected one however it was too small to take a biopsy at the time). I was upset to hear of further growths, however, the one insulinoma was treated with a new treatment which involved injecting the insulinoma with alcohol to try to break it down. This treatment is still in its research stage, however for me it was a success. I had a course of two injections via an endoscopy. The side effects afterwards were uncomfortable however, the fact that I didn't need open surgery again was a big relief and it meant recovery time was a matter of days.

I have a fantastic team who look after me at the hospital and I know that if I ever have any problems I can get in touch with them without hesitation. I now have an annual check up when my bloods are checked along with an endoscopic ultrasound to view my pancreas as this seems to be my troublesome area. My consultant wants to ensure that I hold onto my pancreas for as long as possible by trying to deal with any new growths as quickly as possible. There is no guarantee that I will not continue to get insulinomas. However, regular checks keep me positive.

At present I am awaiting an endoscopy in December 2012 as there are small growths located on my pancreas however, they have been classed as non-specific therefore my fingers are well and truly crossed!!

Life has its ups and downs living with MEN1, it's important to try not to worry about what may happen and instead enjoy life to the full, dealing with things as and when they arise. It's amazing the difference steady blood sugar levels have on you and I'm just thankful to my fantastic husband, family and friends who continually support me along this journey, thank you to each and every one of you.

## **Alison Smith, UK - Benign Insulinoma, 2009**

*"I told my consultant I was ten weeks pregnant... they had never known anybody with an insulinoma to become pregnant"*

My eldest daughter Emilia was born on Thursday 9th August 2007 and until four weeks after her birth, I had never experienced a single symptom of an insulinoma and more to the point had never even heard of one!!!

The very first time I had a hypo, I hadn't eaten breakfast or lunch and had gone shopping with my husband and four week old baby. My mum had invited us round for dinner and when I arrived she thought I was drunk! Apparently I was slurring and talking absolute rubbish. I have no recollection of arriving at her house or how I behaved. During dinner I couldn't answer anybody's questions and kept staring into space. Once I had finished dinner, I returned back to my 'normal' self and everybody who had been sitting round the table told me how I had behaved. I was obviously shocked but we all put it down to me having a new born baby and not eating.

About a month later I was invited to a friend's house for lunch. I actually drove to another friend's house and parked there in order to walk with our prams and new born babies the rest of the way. I have no recollection of walking there, carrying our prams up the steps to her flat, or what happened once I arrived. It wasn't until my friend made me a cup of tea that I realised I had had another 'funny turn' as I called it! Luckily I have always taken three sugars in my tea, so the instant sugar rush had brought me out of my hypo. Both friends then informed me that they thought they had offended me as I suddenly stopped talking to them and when they asked me if my then ten week old baby needed her bottle, I just couldn't answer and was slurring my words. They advised me to go to the doctors but I simply put it down again to not eating breakfast and walking such a long way.

Over the next few months I had a few more 'funny turns' but always caught them in time by eating something sugary and being revived by additional food.

When my daughter was 11 months, my husband and I attended a wedding ceremony that started at 10am. I actually did have breakfast but the wedding buffet didn't appear until 4pm! I then had a couple of glasses of wine but was fine. It was a lovely hot day so I ended up talking to some friends outside. That is the last thing I remember. Apparently a friend went to find my husband as I suddenly appeared very drunk, was slurring and couldn't stand up properly. He then took me home and put me to bed.

The next thing I knew, I woke up in bed to find three ambulance crew standing around me and a drip in my arm. I was unable to speak and was so disoriented as I had no clue as to what had happened to me.

My husband's side of the story is that he woke up to find me having a seizure in bed so called an ambulance. When they arrived I was unconscious and had a blood sugar level of 1.2. They



rushed me to A&E where I spent the day on a glucose drip and undergoing tests. I remained in hospital for a week where they monitored my blood sugars and I underwent a 72 hour fasting test which had to be stopped after four hours as my blood sugar level was dangerously low. The first time I ever heard the word 'insulinoma' was during this week. After having an ultrasound and being given the all clear, I was led to understand that an insulinoma was so rare, the chances of me having one were extremely slim!!!

I suffered constant hypos during the next few months and underwent numerous tests to rule out an insulinoma. I was meant to return back to work after my daughter's first birthday but was then signed off work sick by my GP as I was feeling so unwell. I was told to surrender my driving licence to the DVLA as I was deemed a danger on the road after episodes of becoming unconscious with no real warnings.

When my daughter was 15 months, I went to see my consultant who told me my tests were inconclusive so he was referring me to an endocrinologist at Charring Cross Hospital just to be on the safe side. I told him that my husband and I wanted to try for another baby and he told me to go ahead as there was just no chance that what I had was serious.

Two months later I went to see the consultant at Charring Cross who told me that after going through my test results, I did have an insulinoma! I then informed her that I was actually 10 weeks pregnant and then the whole dynamic changed. According to the endo team, they had never actually known anybody with an insulinoma to become pregnant!

I was then referred to Hammersmith Hospital in January 2009 and under their constant care, as well as the care of the midwives at Queen Charlotte's Hospital which is actually located next door to Hammersmith Hospital.

I found it extremely tiring travelling up to London constantly for endo appointments and midwife appointments. I had to get two buses and two trains to the hospital each time I went, which was very often during my pregnancy. The endo team and midwives team worked together well and I always had one appointment to see them all in one place, which made my day so much easier. I remember scaring my midwife when I had been sitting talking to her for ages and she decided to test my blood sugar which was 1.2! She ran out of the room and returned with a bottle of Lucozade and ordered me to drink the lot!!! She just couldn't believe that I was fine and not unconscious with such a low blood sugar!

My pregnancy went smoothly and I actually felt better than I had done in months. I was given guar gum, a natural plant ingredient, which was mixed with drinks before every meal to control my sugar levels.

It was agreed by the 'medical team' assembled to look after me, that I would be induced as it would be too dangerous to allow me to go into labour naturally. I was admitted to Queen Charlotte's on Monday 3rd August, ten days before my due date. When examined I was actually

found to be 3-4cm dilated so my waters were broken and labour began. I was immediately placed on a glucose drip and told to eat when I could. The team wanted me to have a quick birth so that my sugar levels wouldn't drop too low. I was put on a hormone drip to speed things up. Isabella was born 4 hours 19 minutes after my waters were broken at 12:19am on Tuesday 4th August perfectly fine.

I stayed in hospital for three days where the endo team and midwives monitored my sugar levels. No longer being pregnant, my sugar levels plummeted and my hypos began again. Not easy to deal with when I returned home with a new born baby and a couple of days after, Emilia celebrated her second birthday.

I think I managed quite well looking after a new born and two year old in the first few weeks. I remember walking to preschool (I was still not allowed to drive) pushing a double buggy and eating sugar lumps just to get me there!

When Isabella was ten weeks old, I had a severe hypo. I had been cleaning but didn't think I had overdone it at all. My mum and dad had been with me all day and left at 4pm, putting Isabella to bed in her cot before they did. I sat down with Emilia to watch TV and my next recollection is opening my eyes and seeing two ambulance men in my lounge!

My husband had come home to find the house in darkness and could hear Isabella crying in her cot upstairs. Emilia was sitting in the dark watching TV and I was unconscious on the sofa! After I had suffered severe hypos, I was given a glucose injection which I had to keep in the fridge in case of emergency. My husband was shown how to administer it on me but had only practiced on an orange! When he had found me unconscious, he immediately injected me but nothing happened. He then called an ambulance. When the first response man arrived, he checked the syringe and informed Paul that there was already water in it which he should have emptied and then drawn up the glucose and injected me. Anyway after putting me on a glucose drip, I came round but refused to go to hospital as there was no need, I knew what had happened!

In the months that passed, I underwent numerous tests, ultrasounds, CT and MRI scans to locate the insulinoma's exact location.

On 20th May 2010 when Isabella was nine months old I underwent the major operation to remove my insulinoma. I wasn't nervous at all, more excited that my nightmare would be over! I was told that there was a slim chance that I might have keyhole surgery but when I awoke in recovery I was told that I had had open surgery. I was given an epidural and morphine button as pain relief.

The first few days were a blur as I was heavily dosed up with pain relief.

On the fourth day the surgeon came to look at my wound which measured 30cm with over 50 staples. I was told there was an infection in my wound so four staples were removed in order to

swab the area. Little did I know that removing staples so early after an operation meant that I was left with an open wound. I stayed in hospital for a week.

I left with a gaping open wound and once home a district nurse had to come in daily to clean, pack and dress the wound. After ten days I went to my GP's surgery to have the remaining staples removed. The nurse had to call in a colleague when she got to the end four staples as she couldn't get them out by herself. She told me she thought the staple gun had misfired as the staples were all intertwined. In the end she had to remove them in bits. I was on the couch for an hour and a half. Once the staples were removed the nurse told me that my wound had opened up at the other end. She called my GP in to have a look and he said he would contact my surgeon as he could see down to my internal stitches and thought I should go back to Hammersmith to have my wound re-stitched.

My surgeon said re-stitching was not an option, so both sides of my wound were cleaned, packed and dressed daily until I was feeling ok to travel back up to London to see him. Two weeks later and feeling like death, my husband drove me to Hammersmith. My surgeon said I had been very unlucky both with the infection and the staples. He decided to remove my internal stitches and dead tissue for my comfort. I didn't feel a thing and had no pain relief as the wound was so deep; it was numb from the dead tissue.

A district nurse came daily from May until September. I couldn't do anything for myself, was constantly sick and in pain. I couldn't be left alone, so somebody, mostly my mum or sister had to come to look after me and the girls when my husband was at work. He then took over everything with the girls, cooked dinner and looked after us all when he returned home from work. I slept on the sofa for the first 4 weeks home, as I just couldn't climb up the stairs to my bedroom.

I watched Isabella learn to crawl, eat solids and walk all from the comfort of my sofa. I was told to do nothing in order to aid my wound's recovery and not to lift anything remotely heavy, not even a kettle let alone a baby. I didn't pick Isabella up from the 19th May (day before my operation) until mid-November when my GP finally said my wound was strong enough not to split open. Six months without picking up your baby is a long time and I feel I missed out on the whole first year of her life!

2010 was an awful, awful year but 2011 was a different story. My husband and I went on holiday in March 2011 and had a fantastic time! I was even able to get travel insurance as before I was deemed uninsurable as an insulinoma is such a rare thing.

Now two years on I feel great. I have two lovely girls who are now five years and three years old! Both are true daddy's girls which I think really comes from him having to look after them for so long as I was just too ill to do it myself.

Without my husband, girls, mum, sister and family I would not have got through such a terrible four years. I think myself lucky everyday but live with the dread and uncertainty of the possibility of finding another insulinoma.

## **Paula Davis, Bristol, UK - Malignant Insulinoma, 2009**

*“Being isolated in a lead lined room for 24-28 hours...”*

In February 2008, my housemate found me running between my home and my neighbour's house seeming vague and saying I had to find the binoculars. I was in fact cat sitting for my neighbour. This was followed by an incident of answering the door and just laughing at the poor man stood there trying to engage me in changing my power supplier. Following this, I flaked out and went to sleep for a few hours.

Another incident that month, which all occurred in the evenings following a strenuous walk home from work, I was reading my e-mails and ended up “getting lost in my computer”. It was like being in the middle of a computer game but only for real. The other strong memory I have, is trying to send text messages from my mobile, which I do a lot, and realising at that point, I did not seem able to work my phone. Most of these episodes ended with a tingling mouth and eating to bring myself back to normal. On reflection, I can now see incidents that occurred during 2007 that perhaps were indicating the onset of this illness.

These episodes put together made me make an appointment with my GP during March, I had blood tests for many things but all came back negative, the nurse thought I might be diabetic at one point, as I appeared to have low blood sugar after a fasting blood test. The one area I was aware needed attention was my diet, which was not always good, I was also aware I had been putting on weight for no real reason over the last six months or so. As everything came back normal, I was referred to a neurologist.

In April 2008, I never got to that appointment. At the end of April, I was shopping in Broadmead in Bristol and I remember feeling hungry, as I had skipped lunch, so went to get a sandwich from M&S, they did not have any. I can remember feeling as if I was falling backwards and someone asking if I needed help, I thought I was OK. Next thing was I woke up in an ambulance at 6.15p.m. several hours since I remembered. I had very low blood sugar and was told by the very kind medics I had had a hypo episode, the staff in Tesco had seen me wandering around appearing odd, sat me down and had given me a drink but I had then passed out. I wanted to get up and go home, I had only been shopping! But ended up in A&E where the doctor informed me that due to very low blood sugar I would be kept in. That started two weeks of being in hospital.

I had always been fit, taken regular exercise, walked a couple of miles a day, did not smoke and drank low levels of alcohol. I generally looked after myself, I held a responsible, sometimes, stressful full time job.

Whilst in hospital, a large number of tests followed. It started with two hourly blood sugar tests 24 hours a day. Each test showed I had low blood sugar, especially overnight. After several days, I met Dr Karin Bradley and her team; she is an endocrinologist at the Bristol Royal Infirmary. Quite early on, she told me she thought I had an insulinoma; a friend went off to explore the

internet for me. Everything seemed to indicate it was rare, usually benign and could easily be removed via surgery and noted that it could be difficult to locate. At this point, I was scared of what was happening to me but I did not feel overwhelmed.

I left hospital taking Diazoxide, which is a drug that acts on inhibiting the release of insulin from the pancreas but does have many side effects. As an outpatient I then had a MRI scan and an endoscopy – which indicated a 5cm growth on my pancreas which was obscuring the vein to the spleen, CT scan and finally a 111 Octreotide scan. Following all this, I was told that they had identified the insulinoma on my pancreas and an area of concern on my liver, plus an area in my chest. Following this, I was told I would also need my spleen removed as the insulinoma was obscuring the vein feeding it.

During this period I was taking my own blood sugar level tests at home, they went from a low of 2.1 in the mornings to a high of 19.7 in the evenings. It was a difficult time, as I had to eat every two hours, I had to be constantly aware of my sugar levels and I always carried a bottle of Lucozade to drink and some biscuits to eat with me. Generally, I felt low, exhausted and depressed. On reflection, I also realise that over exertion and stress caused a significant drop in blood sugar levels. During this time, I was in regular contact with the endocrinology team via email and 'phone, which was extremely helpful.

In July 2008 I went into hospital for the operation. Mr Ian Pope, who is a pancreatic specialist, and his team operated on me, this went very well; I went in to the preparation area at 8.30 am and woke up at 2.00 pm in the recovery area. I was then transferred to the high dependency ward where I stayed for three days. During this period, I had intense one to one nursing and every monitor was checked and attended to as required. From there, I moved onto a general medical ward where I continued to have intensive care for a further 48 hours prior to be moved on to the ward proper. I then had four days during which time I slowly started to do things for myself, which was great. Ever since waking up after the operation, I realised I felt so much more alert and positive. The low blood sugar had stopped which in itself was a wonderful feeling.

I was told after the operation that my liver was clear and they had removed approximately two thirds of my pancreas, along with gall bladder and spleen as planned.

The recovery period from this operation is three months of rest and recuperation so I now need to rest and learn to do nothing.

In August 2008 the results from the surgery took a bit longer to come through than I expected. When they did come through it was quite a shock in that the areas removed had proved to be malignant which in turn means cancer. That it is not in my liver and that it is not a fast growing type is great news. I am scheduled for more scans in September 2008, to check on my progress and for the medical team to revisit the area in my chest. While not good news I feel under the circumstances I am doing pretty well to date. I hope my story helps you understand this rare

illness. With the expert medical team that have supported me through this difficult year, I hope to continue to stay well and positive.

### **Paula Davis - Part two of my story:**

I recovered from my operation and went back to work and thought I was OK. Things started to go wrong again during January 2009, I was starting to get low blood sugars again and I knew what they were this time.

I was on monthly Sandostatin injections for a while but they did not seem to help, so I was back to eating every two hours to keep my blood sugar up again. I was extremely lucky that a dear friend Hob saved my life. I had gone into a severe hypo coma during May, we had spoken earlier in the day and she realised something was wrong. Luckily she came to check on me, getting spare keys from a neighbour. Following that episode, I restarted the Diazoxide drug. My deterioration continued until May 2009 when things came to a head when I was readmitted to hospital as my blood sugar levels were uncontrollable. Back to two hourly blood sugar level tests night and day. Following lots of CT and MRI scans and further blood tests, it was agreed the best way forward was to liaise with Professor Caplin at the Royal Free in London. It was agreed that my medical position warranted the radiolabelled octreotide (RLO) treatment (magic bullet) then only available at the Royal Free.

I finally managed to get home, the worst thing at the time was I had not slept for two weeks. I was now on a high dose of Diazoxide which was causing water retention that was extremely painful and uncomfortable, my weight went up by 12 kilos during this period. At this point I met a wonderful GP, Dr Collyer, who has looked after me in tandem with Dr Bradley at the BRI. Just going up the stairs to the bathroom each day was a major challenge. My life was on hold and once again friends and family came to the rescue as I could not be left alone due to the possibility of low blood sugar coma, which did happen once more.

Following liaison with the Royal Free it was agreed I was a suitable for the RLO treatment. I was shocked to find out I needed approval of the local PCT to pay for the treatment as it was in a different health authority, London. This started a difficult couple of weeks. Luckily I managed to make contact with the person who would co-ordinate my application with the PCT which went to a special meeting for approval as I had missed the monthly meeting. This was supported by my consultant and GP who made the case in writing for the Royal Free treatment.

After two difficult weeks the approval came through and all systems were go. I was booked for my first treatment which is explained as “being isolated in a lead lined room for 24-48 hours during which time a cannula is inserted and radiolabelled octreotide is administered” I must confess I was not looking forward to it.

For my first treatment in July 2009, I was driven up by my lovely brother for a 9am start and started the usual medical checks, update and lots of blood tests. The time came to be shown the

“room”. Well, I was very pleased as it turned out to be a room on the 13<sup>th</sup> floor with windows overlooking Hampstead Heath, which was wonderful, self contained with en suite. It was better than some hotels! The treatment started about 3.30pm which consisted of a trolley with the radioactive medicine being wheeled in and administered intravenously, the consultant administering the treatment stays behind a lead barrier by the door protecting his body. The treatment takes about 30 minutes with a drip prior and post to ensure my kidneys remain clear.

I was given anti sickness drugs but unfortunately they did not work the first time. The period of isolation then starts, relax and let the treatment get to work on destroying the horrid tumours. Following day off for a scan to check the treatment has attached to the tumours. Following the results of this, I was clear to go. So my lovely brother comes back from Bristol (250 miles round trip) to take me home, prepared with a bowl and tissues just in case!

It was wonderful to be home and I just needed to sleep. In fact, a few weeks of sleeping and lots of rest. Over the next two weeks I did wake at night with pains in my chest I can only explain as a feeling of something being eaten away, it was the RLO destroying the tumours. One down two to go! The following two occurred at three month intervals, same procedure each time. I had the final one on 11<sup>th</sup> January 2010, this one took a bit longer to get over and I did have a few weeks of being unable to do anything.

I went back to work late February 2010, this time three days per week, it was a bit more challenging this time as energy levels were not as high. During the nine month treatment period I needed weekly blood tests which my GP arranged to be done at home to avoid visiting areas of infection.

So here I am getting on with my life once again. My medical team have been fantastic; friends and family have all given me the energy and mountains of support to get through it all. Professor Caplin says research so far says this treatment on average gives 2 – 3 years before reoccurrence of the tumours, so fingers crossed.

In September 2010 I underwent an incisional hernia repair operation. I am told this is quite common in operations of this size.

In May 2011 I finally realised that all my energy was now going into just being at work and I was unable to do very much else as all other time was spent just recovering and sleeping! The impact of the treatment depletes your bone marrow which in turn resulted in a low haemoglobin reading of 10.2 which should normally be about 12 – 14.

So, June 2011 I decided to give up work, my finances supported this, not as much money but with what I have been through I decided my life is more important so I took the plunge and decided to make the best of whatever time I have left, hopefully a lot! Wonderful leaving dos commenced with colleagues and my team. I have seen during my journey how many wonderful



caring special friends and family I have around me and I want to enjoy every second of those special moments. I have six monthly CT scans with annual PET scans in London.

Each one of these has shown some growth in the size of the tumours but to date nothing significant enough to require more treatment, plus no hypos! I have also recently been diagnosed with an under active thyroid so I now take Thyroxin which appears to have increased my energy levels.

During one of my appointments with Dr Bradley I asked about wanting to get fit and feel stronger again and was worried about my body not being up to it. She replied “|You will be exercising with no stress now that is different!” That is so true, I now realise looking back on my life I had lived a full life with a highly pressurised job, a busy social life, not a brilliant diet and thought I was doing well. I now live well compared to that period and don’t have stress in my life, if I do it is in small doses, not 24/7.

During the last four years I have also been lucky to receive ongoing support via Penny Brohn Cancer Care who offer complementary cancer care, I have also met some wonderful friends, some who have lost their lives to cancer which makes me even more determined to enjoy mine to the greatest degree possible.

Early in my journey I met Enola Wright who had set up a Facebook page for people with insulinomas. During these past few years we have become firm friends and Enola has been an immense support with her ‘phone calls and visits at those moments when health issues have taken over my life. There are now **137** members on this page who mutually support each other with advice and just a kind word which is so crucial during this journey. Thank you so much Enola, this journey has been made so much easier due to your ongoing support and friendship.

So here I am September 2012 – now 57, still well and really enjoying my life. I have recently commenced a new relationship which feels wonderful and very special.

A malignant insulinoma is not a great diagnosis but remember it is not a death sentence either, just a kick to get on and enjoy life to the full as that is why we are all here isn’t it!

I cannot end there without saying a massive “Thank You” to friends, family and our wonderful NHS, the dedicated doctors, nurses and support staff that have kept me alive over the past four and half years with ongoing medical advancements in the area of insulinomas and for many more years to come too!

## **Enola Wright, Berkshire, UK - Malignant Insulinoma, 2007**

*“Eat a bar of chocolate and take two aspirin”. This was the advice I was given at A&E following a blackout whilst driving.*

I called my best friend to say that I would be with her in 10 minutes. Instead, I ended up two villages away with no idea how I got there. I drove 7 or 8 miles with limited vision, it was a bit like looking through a pinprick in a black piece of cardboard. I recall hitting the curb several times with no vision of anything around me. When I did eventually stop, some 15 minutes later, I was confused; I had double vision, was very shaky, and was weak and extremely disoriented. Somehow, after 20 minutes or so sitting in the car whilst I regained single vision, I managed to drive to my friend's house. She tells me that I arrived there driving very slowly with the car in second gear. She took me to my local A&E where they carried out some basic tests, including a blood glucose test, which had a reading of 2.6. Then the advice they gave me was to go home, eat a bar of chocolate and take a couple of aspirin.

I knew something horrible had happened to me. I wasn't sure what it was, I thought it could be a stroke, and insisted that the A&E doctor referred me to a specialist. A few days later I got an appointment with a cardiologist, who dismissed all of my symptoms out of hand and said, I was in danger of seeing ‘one too many ologists’. This was the same cardiologist who had me on tablets for high blood pressure, which disappeared once my insulinoma was treated. I was also referred, by the A&E doctor, to a neurologist. The neurologist was brilliant, he actually took the time to listen to me. Although he did not think it likely that I had a neurological disorder he sent me for an MRI of my brain. The scan showed that nothing was wrong. He did not dismiss the blackout and continued to ask me quite a few questions about my health leading up to it.

I described the preceding week when I had ‘nearly’ fainted on about four separate occasions whilst at work. I managed to stave off unconsciousness by eating glucose tablets by the dozen and by eating anything sweet. I was also drinking a bottle of Gaviscon every three days or so as I had very painful indigestion. Other symptoms included a weight gain of approximately three stone (over a period of a year), high blood pressure, tingling lips and tongue, shaking hands and light headiness. My tummy was also very distended and swollen which made me look as though I was six months pregnant. Incidentally, the neurologist had a copy of the report from A&E, which contained no mention at all of my glucose reading. I mentioned my glucose level almost in passing as I thought it was low at 2.6. He said immediately that I might have a problem with my pancreas and said that I should see an endocrinologist.

Several weeks later at my appointment with the endocrinologist, she also suspected a problem with my pancreas. Following a 14 hour overnight fast, my glucose level was 2.2 mmol/l, and 2.3 mmol/l on two consecutive days. My insulin levels were also high at 138.6 mIU/L and 145.7 mIU/L respectively (the normal range is up to 24.9) along with an elevated C-peptide of 6.68 ug/litre (the normal range is up to 3.2). A CT scan with 3D reconstruction of the abdomen

revealed a 2.4 x 2.4 mm vascular mass in the tail of the pancreas with some abdominal lymphadenopathy. Other scans followed including an MRI, an endoscopic ultrasound scan and a Gallium 68 Octreotide (which showed the tumour up like a light bulb).

My diagnosis was an insulinoma. Insulinomas are rare neuroendocrine tumors with an incidence estimated at 1 to 4 new cases per million persons per year. 95% of all insulinomas are benign. Mine, unfortunately, turned out to be cancerous with the complication of a metastatic spread to lymph nodes.

In April 2007, I had a distal pancreatectomy to remove the tumour, a splenectomy and eight lymph nodes in the pancreatic area were removed. The operation itself went well but I had major post-op complications, which included a collapsed lung, pneumonia, sub-phrenic abscess, anaphylactic shock, weight loss of 14kg, and to top it all I contracted MRSA. By July 2007, I had made a good recovery and returned to full time work.

In August 2007, follow up scans showed another insulinoma near my adrenal gland and several enlarged lymph nodes. In September 2007, I underwent keyhole surgery to remove the tumour and lymph nodes. Again, this was not without complications, which included a bleed from my kidney and a 3 pint blood transfusion and a pancreatic leak. Following the removal of more of my pancreas I became a type 2 diabetic.

Recovery was slow and painful. By December 2007, I was feeling much better and decided to return to work, full time, in January 2008. My condition is monitored very closely by Dr Caplin at the Royal Free in London and a CT scan during January showed up a suspicious lesion in my liver. This was confirmed by an MRI scan but on this occasion the Gallium 68 Octreotide PET scan did not light up. In April 2008 I underwent radiofrequency ablation (RFA) to remove the lesion on my liver.

In July 2008 I managed to get an incisional hernia at the site of my first operation. The procedure involved opening up the entire scar. I also had a tumour like mass removed from my abdomen, which fortunately turned out to be benign. Recovery again was very slow and painful.

I have three grown up children aged 26, 22 and 20. Despite a diagnosis of cancer I try and stay positive about the future. I continue to be closely monitored by Dr Caplin, and my aim going forward is to help raise awareness of insulinomas.

## **Angie Phillips, Woodford Green Essex UK - Benign Insulinoma, 2006**

*Small but perfectly formed to threaten your life!*

*"I am suffering from an insulinoma (I am not a ferret or a dog)...."*

That was my first post that started the ball rolling on a forum devoted to insulinoma and Whipple triad surgery. (<http://www.unboundedmedicine.com/2006/01/15/insulinoma-whipple-triad-surgery/comment-page-17/#comment-543565>)

It was to become a life line for myself and several dozen people, and still is.

It was May 2006, I had pulled my back somehow, not that unusual but I hadn't remembered doing much to cause the extreme pain I found myself in. It was very odd as the following day the pain had improved so much it was almost like I had imagined it, however I went to see my doctor. I truly believe now this was fate throwing its hands in the air and insisting that I couldn't ignore my health any longer.

At the same time as discussing my back pain I requested a blood test as I had a suspicion that I was anaemic due to such tiredness and extreme lethargy, although I explained it as "life" with two kids! My doctor sent me for a blood test, a fasting test.

That was Monday, Wednesday I had a phone call calling me back to the surgery urgently. I wasn't anaemic! We talked through some history, migraines, the severe tiredness, getting up all through the night to go to the toilet, extreme night sweats, and the symptoms I suffered when I didn't eat properly. I would shake violently and lose my speech, and my tongue went 'fizzy'.

My doctor had studied at St Bartholomew's Hospital in London specialising in endocrinology, so this kind of case spiked her interest. She described what she suspected, a tumour that secreted too much insulin but needed to refer to a colleague. Two days later she called me to say her suspicions were confirmed and I needed to come back in.

I was referred to Whipps Cross Hospital, on my request, never making a drama out of a crisis I couldn't see the need to be referred to a London specialist hospital if it wasn't necessary. At my appointment they referred me straight to St Bartholomew's! They gave me a blood monitor to 'play' with while I waited for my appointment!

Within 6 weeks of my initial appointment I had a meeting with Dr Drake (now Professor) and a 72 hour fast was booked.

For years I had just decided I had low blood sugar and that was me! Never realising there were any health risks or that it was a real problem. I remember one particular afternoon before my first child was born, getting out of my car and feeling the need to just collapse and never get up. It was extreme; I had a huge glow in my eyes that distorted my vision, as if I'd been staring at the

sun. I related this to my previous doctor some time later but he put it down to a symptom of my migraines and I had no reason to doubt him.

My blood sugar readings varied but never went over 4 even after eating; normal readings should be 4-6. My levels averaged at 2.3! If you hit as low as 1 you are either in, or about to go into a coma!

When I arrived for my 72 hour fast, they told me the test would stop when my levels hit 2.2! They were a little surprised when I told them that was normal for me!

I lasted 30 hours in the end before they stopped the test. The confusing time occurred when my blood sugar went up rather than down. They weren't sure why, basically my liver kicked in as an emergency reaction producing glucose to 'save my life'!

Anyway after dreaming about digestive biscuits my body reverted to 'normal' and my blood sugar dropped dramatically proving there was an anomaly! I had two hypos hitting lows of 1.9 and 1.6.

One of the difficult hurdles in being diagnosed with insulinoma is that it's so rare not many doctors come across it. 1 in 250,000 people get an insulinoma. The reason Whipps Cross did not want to do the 72 hr fast was that their nurses were not trained to cope with it.

I was then sent for a MRI and CT scan. It was confirmed that I had a 1.5 cm tumour in the head of my pancreas, but I was also booked in for a calcium catheter investigation. I was sent home with the wonderful Diazoxide tablets which were supposed to help control the insulin; I was soon on diuretics to control the oedema, a side effect of the Diazoxide!

It was only around this time that I really understood how ill I was. I started a bit more research, I'm one of those people that like to be informed, not to panic but I needed to know as much as possible. Insulinoma was classed as a life threatening condition. The hypos would get worse and I could lapse into a coma at any time. The tumour could also become malignant. Now I understood why I'd been rushed through the system, I had thought it was a bit odd there had been little waiting time! The opposite of diabetes but with less control. The insulinoma tumour develops from rogue cells in the pancreas that clump together and continuously produce insulin with no inhibitors, totally taking over from the pancreas. Too much insulin removes more and more blood sugar from the body, depleting and exhausting the body's natural supplies! My hair had been getting thinner, my nails wouldn't grow and broke.

Looking back, I think that as the tumour got bigger I coped and my body got used to existing on low levels of sugar. I'd been trying to lose weight - as you do - for years always being 1-2 stone over weight but really struggled. Without realising, this actually had helped me as, if I hadn't been trying and hadn't been following a healthy eating diet, I would have ballooned with the

need to eat and the need to keep my blood sugar levels up. Also the yo-yoing of high to lows would have been far more damaging.

I had started doing more exercise regularly and to be honest I think that is what pushed my body over the edge! I'd get out the pool and shower but by the time I was drying my hair I was drenched in a cold sweat! I'd never been an exercise fan and remember the Rosemary Conley consultant (another of my diet trials) commenting that suddenly during the exercise class I would go completely white and drained... she had been concerned but I never thought about it!

There was very little information on the internet and I spent fruitless hours trying to discover not very much at all. Everything was written by vets about ferrets and dogs, you can imagine the comments from friends!! It wasn't until after the calcium infusion test, an endoscopy and several meetings with my surgeon I discovered the insulinoma forum. By this time I knew that I may have to have a 'Whipple' or complete 'repiping' as I imagined it and I was terrified. My surgeon felt the tumour was too near a vessel and would be very difficult to remove. Finding the forum was odd as no one had posted and being the first I had no expectations! Within a few days more posts appeared and I'd connected with Helen, she had just had her op under the same doctor at Barts and same surgeon, and she lived half an hour away. I felt like crying with relief!

Finding people who understood what I was experiencing was so comforting! One of my so called friends had dismissed my condition as 'nothing' and over dramatic! Luckily, she was the only one and is now no longer a friend! Another was surprised that I had a more serious condition than she did! Overall everyone was great, even if they didn't really understand. Writing on the forum was therapeutic, being able to give advice and ask for help, connecting with people not only in the UK but all across the USA.

My hypos were haphazard but getting worse, I had one incident when I 'lost' time - I don't think I blacked out but it was very disorientating and frightening! The Diazoxide wasn't really doing much and although I had hoped not to have surgery I was told that I was too young to take Diazoxide for the rest of my life as I was already having trouble with it and suffering from the water retention. I had developed hairy toes as well!

Surgery was the only sensible option. It was booked for Feb 2007 at the Royal London Hospital in Whitechapel. Mr Bhattacharya.

I spent Christmas thinking it could be my last (surgery scared me to death) and I was terrified of not seeing my boys grow up! One of my friends however gave me some very harsh but realistic advice - why worry, I wouldn't know anything about it if I did die! Strange but it was comforting - in a way!

I survived my surgery; it was six hours and although I had no real problems from the actual operation, there were several other complications. I actually avoided an untimely end only the night before when the ward doctor told me they were putting me on an insulin drip! She went away and read my notes when I told her exactly what I thought of that.

The tumour was removed easily, I think they prepare you for the worst! I didn't have to be 're-piped' and suffer a 'Whipple' procedure. Although I had had an epidural for pain relief, that failed after 24 hours and was replaced by a morphine PCA. I was uncomfortable and sore but my blood sugar had reverted to normal immediately.

Being in hospital for two weeks was an experience; it made you very dependent and made you feel a bit helpless but safe. My friends were amazing, Anna came in and helped me bath and wash my hair, Katy came in and blow dried it on occasion to make me feel better. Lesley popped in after work and made me laugh. Without my visitors I would have gone nuts! I met wonderful Nicky in the ward who had had a terrible op on her throat. I laugh now as I remember how we trailed our 'drains' and 'drips' up and down the ward in search of lunch and dinner, hoping to get the better choices than those still bed ridden.

It took a while to get back on my feet, with an 8 inch scar across my middle I told people I had been bitten by a shark - much more interesting! I was very sensitive to some foods and had a lot of stomach pains. I later discovered they had removed my gallbladder which explained much of that! Sleeping was very uncomfortable and the lack of strength in my stomach muscles was frustrating and painful.

Lots of symptoms disappeared which I hadn't even attributed to the insulinoma. My migraines were non-existent, my hair started growing and I had baby hair regrowth. The excess hair and down caused by the Diazoxide disappeared, I had normal toes again!

I could read without the letters jumping around on the page, no middle of the night cold sweats and I didn't have to panic if I hadn't eaten lunch! I tested my blood if I had a 'low' feeling but when I did it was always normal. My extreme stomach pains were eventually cured, a scan had showed no complications, which was good but frustrating, my GP gave me some antacid tablets and it eased almost immediately.

I was very lucky for the insulinoma to be diagnosed so easily and quickly. I spoke to my doctor recently about how rare an insulinoma was, and asked why she went for that diagnosis. She told me it was my extensive history and symptoms that all added together. She sends loads of people for blood tests every day for one reason or another but it is very very rare that the same histories come along.

My doctor picked it up quickly but she had specialised in endocrinology and had been aware of the condition where as many GPs would not have known enough and wouldn't have even considered it. There are so many cases of people who have been turned away and told there is nothing wrong with them – you know your own body and without being a hypochondriac you have to be strong enough to keep fighting for some kind of diagnosis!

Find out as much information as you can, and when you know what the diagnosis is, research it with an open mind. Don't take everything as gospel it may not be relevant to your case but knowing about it makes a difference. The medical profession do their very best but with the best will in the world mistakes can happen. Ask lots of questions, get everything explained and don't be put off if something doesn't seem quite right!

It is easy to forget how ill I was and how ill I could have been. However, I will never forget how lucky I was and the good care I received and the very good friends, those I made at the time, those I've made since and those who cared enough about me to get me through it all.



## Wendy Remnant, UK - Benign Insulinoma, 2006

*"I just needed something sweet"*

I really don't know when it all started but I think that it might have been as long ago as 2002. We would have a meal, with a cup of tea afterwards. I would help myself to a small cake or biscuit saying 'I just needed something sweet'. I haven't had sugar in drinks since I was 7 but having done a Weight Watchers diet once, sugar free drinks were the norm for me. It was later on that my husband said that it was a habit of mine – a very regular habit with a reason I didn't know about.

By 2004 I first started noticing things. Bad headaches, especially after one alcoholic drink the previous evening, lethargy, lack of concentration and awareness. I just put it down to working for myself and running around after the children and my age.

It was Tuesday 7<sup>th</sup> June 2005 when I knew I had a problem. I had an early lunch – sandwich and yoghurt probably and left home to drive to do a bridal makeup rehearsal booked for 2pm about 40 minutes from home. The rehearsal took longer than planned and I had a drink of water whilst I was there. I left around 6pm ish – I don't really know what the time was. I wasn't feeling too good and I knew I needed to eat something. I had an empty vagueness around me. Something was wrong. I knew of a shop nearby and drove there. I sat in the car for a short while then went into the shop. I knew I had to eat but I didn't know what to do in the shop. There were people there and it was all very confusing to me. It wasn't a panic attack just confusion. I went back to the car and sat there again trying to make myself feel 'right'. I had a pack of tic-tacs in the car and just sat there eating them. I decided I had to get home. I 'phoned my husband to say I was on my way and he said he would get dinner.

That drive was so long. I drove slowly as I knew I wasn't concentrating. I remember at one point wondering what the gear stick was for. I really have no idea how I managed that journey without having an accident. Fortunately I got home. The family had just sat down to eat. It was strange but I felt my body returning to normal as I ate. It was only a salad but the effect on my body was amazing.

The first thing I did the next day was to look up my 'symptoms' on the internet. By this time I thought absolutely the worst. Early stages of dementia? Brain tumour? Alzheimer's? I used Google and it took ages to sift through the sites but I didn't come up with an answer.

Next day I 'phoned my GP surgery. I was given an appointment with a locum GP. At the appointment I carefully explained what had happened to me on the Tuesday. The GP listened and at the end took a reading of my blood sugar which showed a level of 4. 'Don't worry, it's just a one off thing, you're not diabetic'. I was dismissed. BUT I know my body.

Back to Google.....

It took time but eventually I found a site related to diabetes. I knew it wasn't diabetes as I have a diabetic niece but scrolling through I found out about hypoglycaemia. It all seemed to fit. There was a long list of symptoms and I could relate to some of these. There was also a list of food/drinks that would help when an attack was imminent. I printed the whole lot. I mentioned to my husband about me and my experiences, and showed him the print off from the diabetic website. I took to carrying a pot of raisins with me now but I don't think either of us were quite aware of what was going on.

Time passed and I thought perhaps I was OK. Then there would be the odd time when I knew I was 'losing it' – vagueness would descend over me. We went on holiday to Spain in August and I recall one day feeling quite distressed. My kids got me some orange juice. I'm not sure that they all then understood what was going on – I didn't know myself.

**Monday 31<sup>st</sup> October 2005.** I remember a sense of movement, everything was bright white and I could hear a voice. The voice became clearer – a man was asking me what day it was and if I knew the date. I remember thinking it was a silly question but answered Monday – it was Halloween, 31<sup>st</sup> October. The whiteness dissolved into reality – the man was a paramedic and the sense of movement I felt was because I was in an ambulance heading to hospital in Canterbury.

It transpired that my husband had woken up that morning as the phone was ringing in our bedroom. It must have been around 8am. I usually answer the phone but I was just lying there – eyes open, apparently awake. Both he and the children could get no sense out of me – I said something like 'Go away, I'm OK'. I obviously wasn't. However, as I appeared awake they didn't act with any urgency. My husband, not knowing what was wrong or what to do 'phoned the GP surgery. They couldn't get a doctor out until after 12 noon. Apparently the children were quite distressed and so he phoned 999 for an ambulance. The paramedics arrived and immediately diagnosed a hypo attack and did whatever was necessary for me. **A blood sugar reading showed 1.1.**

In the hospital emergency treatment centre I learned that the cause was two glasses of wine the night before. We had popped out to the pub – a very rare event - when we got home I had a large glass of squash – low sugar squash. This was my big mistake but I didn't realise. The wine reacted badly with my body, pushing my blood sugars right down to floor level.

An appointment was set up for me to see a doctor at the hospital as a follow up on 9<sup>th</sup> November. He immediately referred me to Dr Michael Flynn the local endocrinologist. I was given a little advice about alcohol and diet and advised to only have an alcoholic drink with food until I had seen Dr Flynn and was correctly diagnosed. It was also suggested that I get a blood sugar monitor to regularly check my levels.

This I did and I went to my local surgery where a nurse showed me how to use it and to understand the readings.

I still continued to do 'strange' things – most of which I did not tell anyone about – I remember one day sitting on the bed wondering what to do with a bra and how I wore it. I would go places and have virtually no recollection of it shortly after. I kept eating regularly to try and push the blood sugars up and was steadily putting on weight.

9<sup>th</sup> January 2006 – first appt with Dr Flynn. He initially diagnosed hypoglycaemia and arranged for me to have a series of tests. He also confirmed that I needed to constantly check my blood sugar levels and have a snack when they were dropping.

17<sup>th</sup> January – First test was a CT scan – the results subsequently showed nothing. On 20<sup>th</sup> March Dr Flynn prescribed Diazoxide as my blood sugar levels were constantly low. However, I subsequently stopped taking this as I suffered distressing side effects and the drug had no effect on my symptoms.

There followed a succession of visits to the hospital, still under Dr Flynn, for further (calcitonin) blood tests, another CT scan and finally on 22<sup>nd</sup> May hospital, Dr Flynn, believing that I had developed an insulinoma, referred me to Mr Richard Collins at the Kent and Canterbury Hospital. Mr Collins had experience of my condition.

19<sup>th</sup> June – I first met Mr Collins who heard my story and said he felt the insulinoma diagnosis was correct. An MRI scan was set up to try and locate the exact site of the lump. This was done on 4<sup>th</sup> July but nothing showed on the scans. I was booked in for surgery on 31<sup>st</sup> July. It was explained to me that depending on the site of the lump I might have to undergo a Whipple's procedure which apparently had the trainee doctors queuing up to watch!!

My personal thoughts were that my life would start again when I woke up.

I was lucky – the insulinoma was towards the 'tail' of the pancreas. They used an ultrasound during surgery to trace it. It weighed 0.8 of a gram and was roughly the size of a pea.

Eight days in hospital and I came home just over a stone lighter and feeling normal again. When I went for my post-op check Mr Collins reaction was – 'Don't you look good!'

Mr Collins has since retired but I am so grateful for the large scar he left me with - a small price to pay!

## **Kim, Canada - Benign Insulinoma, 2006**

*“My lips would tingle, hands started to shake and then I would feel like I was in a dream”.*

I would have to say the first time that I can remember anything about my symptoms was when my son was two years old, it was Christmas day 1997 and he was playing around the Xmas tree and it toppled over on him. I got up to help but all I could do was just stare! I knew what was happening but was unable to do anything about it. I was so disoriented. My wife was angry at me for not helping.

That was the beginning!

Over the years it would happen again and again. I would start to sweat, my lips would tingle, hands started to shake. And then I would feel like I was in a dream. Everyone would say “Eat something, you'll feel better”. So I ate, and I did feel better.

Yeah I was gaining weight-- I used to be very outgoing, always working on something, going somewhere, but now it was getting harder. Decided to start jogging, made it to the end of the road and had to come back and chug down two cokes. Thought it was me - thought I was just lazy!

A few more years went by...

Being a man, a stubborn one at that, I kept telling myself everything was ok. Sometimes my wife would be talking to me and I could see her lips move but I was somewhere else. The family and I went on vacation and after a day of walking around we decided to stop for lunch at a fast food restaurant. At the counter, my wife asked me what I wanted, I could see the menu on the wall, but I could not read it. I did not know where I was. It was all so confusing. I sat down and they ordered for me. It was time to see a doctor.

My family doctor was on vacation for the summer so I saw his replacement. I went for a fasting glucose test and it came back 1.2 mmol/l He said I was hypoglycaemic and should take glucose tablets if I was feeling bad. He did not seem too concerned? He also said he would also contact a specialist just to be sure and would call me if anything should come up. A couple of months went by and my wife said I should call to make sure everything was ok. Oops - he forgot to call.

Another month went by and I finally was able to see an internists. Another month went by and I had an ultrasound done. The head physician was called in by the technician to see and he said I think we should send you for a CT scan. I got that appointment in nine days. It was getting kinda scary now.

My internists (a wonderful women - love her) told me I had an insulinoma tumor (never heard that word before) and she explained it all. Of course my first thought was the “C” word, she said if I've had these symptoms for nine years now she didn't think so but of course you never know until surgery and biopsy.

My surgeon (a wonderful woman - love her too) explained to me that they could not pinpoint exactly where the tumor was in my pancreas so they could not do laparoscopic surgery, they had to open me right up. She also told me that because of the same they did not know if they would do a distal pancreatectomy or the Whipple procedure (look that one up).

I had surgery on 2nd February 2006. I woke up after surgery feeling cold and nauseous (threw up a couple of times, once on the nurse, sorry dear). Recovery was slow the first day so they kept me in post-op for quite a while. The nurses tried to get me up after the second day but I was in so much pain I just couldn't and they did not understand this, they said I should have almost no pain. They gave me more pills which didn't work then after one more day they decided to take out my epidural and reposition it. That worked and after fifteen minutes I was up and walking about. After ten days, I was feeling good and wanted to go home so they discharged me. I was quite grateful that I only had the distal pancreatectomy and not the Whipple. Six weeks later I went back to work feeling great!

I went for a follow up to my surgeon's office and she said that my biopsy had revealed not one tumor but three, there was no cancer. With three tumors though, there might be a chance of more in the front part of my pancreas, but time will tell I guess. I am type 2 diabetic now but am managing it well with insulin and pills. I go to the diabetic clinic at the hospital every three months.

Life is good and now I do everything I did ten years ago without all the "fuzzies" as my wife calls them. My only regret is that I did not get myself checked sooner as I could have avoided all that was happening during that time. It was a wild ride but one I care not to take again.

## **Graham Carter, Tewksbury, UK - Benign Insulinoma**

*"I apparently became 'not a nice person...., picking fights with family, occasionally going 'off the handle'"*

Insulinoma, directly, and indirectly has been part of my life for the past 20 years.

Until 2006 I led a relatively normal life, I had a high pressure executive job in London and lived 100 miles away in the Cotswolds, either staying in London in the week or travelling daily. Prior to 2006 various 'incidents' occurred, which on reflection and discussion with various eminent medical professionals, coupled with stats from other cases and my own recollection of events have enabled me to paint a picture of my life leading up to and after the discovery of my insulinoma.

Working in London, sometimes travelling 1,000-1,500 miles a week, and after seven years in my role it became apparent to human resources that I was 'tiring' and had lost the enthusiasm for the job, and the organisation asked me to consider relocating to London with my then wife and family, which we did indeed consider, but eventually rejected, so a year later I left the job and headed home to a new role in the Cotswolds.

During the latter two years of my job in London, yes, I did change, although I was not directly aware of it – I was more tired (*I fell asleep on the train twice and missed my station*), I was sometimes aggressive in debates in meetings, and occasionally 'throwing wobblers' over trivial matters. Whilst driving I frequently needed to stop off somewhere for half an hour or more before being able to continue the journey (*this alone is not unusual, many company representatives can be seen 'napping' in a layby or service stations*). So, on its own not something which can be used as a diagnosis.

In my home life I apparently became 'not a nice person' to quote my ex-wife, picking fights with family, occasionally going 'off the handle' over something trivial.

I can recall one evening when visiting in hospital, as I entered casualty I felt 'weird' and collapsed into an empty wheelchair, promptly to be ejected from the chair bodily by a passing porter and being kicked out of casualty for being 'drunk'!

Various collapses happened, in town, in Tesco's, at home and once in my clinic and at work elsewhere which was the turning point in discovering the insulinoma. But, before moving on to the discovery of insulinoma I was referred to the diabetic clinic, because insulinoma collapses are like hypo-diabetic collapses, sudden and unexpected! I was given a 'GlucaGen HypoKit', a 1mg ready filled syringe for injection if needed, strangely though it was never used... I also started taking regular blood samples, the normal reading should be 4.5 - 6.0, my bloods were very often much lower, in fact the lowest I ever recorded was 1.4.

Over the coming months various other collapses occurred, including the one in August 2006 whilst at work, which was to be the start of the path of discovery to my insulinoma. On being loaded into the ambulance the crew took obs as usual, and declared I seemed fit and fine but they would take me to casualty as a precaution. As they got to the airport entrance they turned left, and not right. ‘Mmm’ I thought, at the next traffic lights they turned left again and then I realised I was not heading to my town, but one just 8 miles away... On asking why I was being taken there I was told they were a crew from that town!

On arrival at casualty I was eventually seen by the casualty consultant, who had spent some time pondering my case, and looking at my notes from various other incidents and also the diabetic clinic reports. He concluded there and then that I may be suffering from something much rarer. He told me he had been a diabetic all his life since a kid, so was very aware of diabetes, and that in fact it was one of his specialisms.

I was referred by the casualty consultant to a professor and his team at Bristol Royal Infirmary, where I underwent an endoscopic examination in November 2006, confirming the casualty consultant’s possible diagnosis. I had an insulinoma but needed to be referred again to the leading UK endocrinology team at London’s Hammersmith Hospital.

Under the direction of a professor and various senior consultants, I spent the next six months (*February 2007 – September 2007*) undergoing various tests and investigations, including yet another two endoscopic examinations, two MRI scans, as well as PET, Gamma and CT scans of various types, at one point I think I actually glowed in the dark because of the volume of radioactive material being injected for imaging purposes. I also had to have two angiogram calcium stimulating tests, also called an intra-arterial calcium stimulation test, the second one because the first one failed.

By September 2007 I was ready for the operation, both personally and also because the medical team wanted to move quickly before the insulinoma became worse, or even inoperable? On the day of the operation I walked to theatre, waking later after a 7.5 hour operation by four specialist consultants, including one flown especially from France. Surrounded by family as they thought I may not survive the night I was heavily sedated, I had tubes out of my stomach, up my nose, out of my neck, plus a catheter and usual Venflon butterflies inserted in my hands/arm for post-op medication/antibiotics etc.

I was told I would be in hospital for probably three to four weeks, but after a very rapid recovery I returned home just ten days later, and that would be the start of a new journey.

Some weeks later, having been to the post-op clinic at Hammersmith Hospital and been given a good prognosis (*I was healing really well, and the insulinoma had been tested, was benign and non-cancerous*). Out of the blue I became ill, collapsing as before, this time though in terrible, terrible pain, sweating profusely, abdominal cramps that lasted 45 minutes to an hour, it was

awful. I was rushed on several occasions to the local casualty department as the concern was I may be suffering from pancreatitis from the insulinoma resection.

After about three or four unexpected visits to casualty, I had an ultrasound scan, which uncovered that I had gallstones, so for another year I had to endure further unexpected bouts of severe pain before the operation site could be re-opened. So once again, diet was in check, making sure I did not eat fatty foods, drink milk, have butter etc.

July 2009 saw my return yet again to London's Hammersmith Hospital for a cholecystectomy (*gall bladder removal*) by the same consultant that did the original insulinoma operation in 2007. Having had my gall bladder removed I found myself with bouts of unexpected hiccups after drinking or eating, luckily this disappeared as quickly as they appeared, just one complication sometimes of gall bladder removal!

How did I get gallstones after the insulinoma, simple – during the original operation in 2007 for insulinoma they tried to perform the procedure by keyhole surgery, but due to the location of the insulinoma they had to convert to open resection, which meant removal of the stomach and gall bladder to get access to the pancreas, hence disturbing the gallstones...

Keen to get back to work I think possibly I returned too early, and ended up a few months later with a post-op hernia, my 'Alien' as I nick-named the sticking out lump, so yet again in 2010 I revisited my now favourite Hammy Hospital for a post-op hernia repair. Again, by full open surgery I had the hernia site repaired with insertion of gauze to bind and hold the 8" incision, made originally in 2006, together.

Summer 2011 has seen me finally discharged from Hammersmith Hospital after five years, 100% fit and well, with a caveat that if ever I feel unwell, or unsure I have a 'hotline' to the endocrinology team at London's Hammersmith Hospital for life.

I still occasionally check bloods, curiosity I suppose, but unlike removal of a gall bladder, or appendix whatever, we cannot survive without a pancreas, it produces numerous other substances apart from insulin which all interact with the body organs to give us life. I will always be grateful to the casualty consultant at Gloucester Royal Hospital who started the journey of discovery of my insulinoma, without his 'intuition', his forethought I may now be floating on clouds.

I must also record my utter, utter faith in Hammersmith Hospital, London, it may look old and decrepit by today's standards, but the level of patient care by the consultants, doctors, nurses and others is first class and second to none, something that you need when you are a very, very rare case. In 2006, the year before my operation Hammersmith Hospital had just 14 cases from the UK referred to them, scary, but all survived...



Interestingly, the comment from the professor in charge of endocrinology at Hammersmith was that they are usually slow growing tumours and they rarely spread. The size of my insulinoma indicated that it had probably been growing for about 15 years pre-op, coupled with the 5 years post-op complications mean I have lived with insulinoma and its associated effects for nigh on 20 years, and survived to be 100% fit and well today.

## **Janet Bennett, Tamworth, UK - Benign Insulinomas, 1975 and 1995**

*“... I was diagnosed with epilepsy and given phenobarbitone”*

It was 9<sup>th</sup> March 1974, my 17<sup>th</sup> birthday, and I was a bit naughty! I had a couple of drinks with my boyfriend, the first alcohol I ever tasted; but from that moment my life was never the same again. I had two Bacardi and cokes, but what I was unaware of was the presence of an insulinoma, I went to bed that evening around 11 o'clock, but at 5am my mom awoke to me having a seizure. I did not regain consciousness and it was thought I was just sleeping. However, my mother tried to wake me in the morning but I was in a coma. My dad came to visit as she could not rouse me and it was his nursing experience which led him to believe I was hypo. He put sugar on my lips and eventually I started to wake up. It was 4 o'clock the following afternoon when the doctor arrived and it was agreed I would have to have tests for the explanation.

They ran fasting blood sugars and an investigation for epilepsy; despite a normal E.E.G. I was diagnosed with epilepsy and given phenobarbitone. Several weeks later I had new symptoms of going into episodes of not knowing where I was, collapsing and generally being unable to go many days without some very strange episodes. My parents knew that sugar put me right but had no idea why. I was admitted to hospital and sent for more neurological testing; brain scans etc. I thought I was going mad; I was unable to keep up with my ambition to be a professional French horn player, despite my winning a place as a junior student at Birmingham School of Music, I had too many black outs. My follow up appointment after being discharged from hospital yet again, was attended by my father who pleaded with the neurologist to look closer at the blood sugar levels as he felt this was the cause of my fits and comas.

I had more fasting tests and was sent home. What I was unaware of was that these tests had to be sent to Innsbruck for the insulin levels, we had not heard of insulinoma and in those days that was the nearest place for testing insulin levels.

Almost a year after the first seizure and still not diagnosed I fell into a dangerous coma on a bus and when reaching the terminus the “clippie” (bus conductress) sent for an ambulance. I was taken to the Royal Worcester Infirmary where they had no idea why I was unconscious. A police man arrived on my mom's doorstep to ask to ring this hospital. She told them that sugar would bring me round and after testing my blood sugar I was given intravenous injection. My blood sugar was recorded at 0.4 which is so low. The following day it was revealed the tests were back from Innsbruck and I was told I had a tumour on my pancreas secreting insulin! I was transferred to Dudley Road Hospital Birmingham and everything happened really quickly after that. I had an arteriogram which revealed the insulinoma. A few days later I had a third of my pancreas removed, my spleen and appendix. A huge op for an almost 18 year old, but what a relief to know I was not mad. I so thought I had lost my mind.

All went really well in my life after that brush with death until 1995. I was walking up the beach in Gran Canaria and all of a sudden the blackness came back; I knew I was dangerously hypo and tried to fight passing out. I could not believe it was back. Over the following months I had tests again which revealed insulinoma recurrence! How could my life be tipped back upside down again! I did see a surgeon but together it was decided the op would be really difficult because of my previous surgery. I decided to try the Diazoxide. I have been taking 600 mg of this drug now since 1996 and I have to say it's my life line. I do still have hypos, but they are controlled with the Diazoxide and careful dieting. After so much of my life with this disease at the age of 56 it is now just part of me. I get on with life, taking pills and always having to think about how to eat, when to eat, what to eat and take my tablets. But wow I am so blessed to be here; I play my piano every day, look at my family and thank God for Diazoxide! Yes I have huge scars, both physical and emotional, but after reading the stories of others it has helped me so much to know I am not alone. It was years before I ever managed to speak to others who have battled with the same.

Insulinoma is just part of me, but there is so much more to enjoy!

## **Part 2**

### **What are insulinomas?**

Insulinomas are a rare type of functional neuroendocrine tumour usually found in the pancreas. They are called functional because they produce insulin and cause blood sugar levels to drop, often quite dramatically and in 'episodes'. Even the smallest tumours can cause symptoms connected to hypoglycaemia (the medical term for low blood glucose levels).

The vast majority are benign (with no spread beyond the pancreas). However, 10% are malignant and by the time of diagnosis will have spread elsewhere in the body, usually the liver. The tumours are generally diagnosed in middle age and slightly more women than men are affected. If an insulinoma is suspected you may also be screened for a genetic condition called MEN1.

## **The role of the pancreas**

In a healthy person the pancreas produces both insulin and glucagon. When blood sugar rises after a meal, beta cells in the pancreas release insulin. The insulin helps sugar from food to enter the blood cells and lowers levels of glucose to normal.

If blood sugar falls too low then alpha cells in the pancreas produce glucagon that triggers the liver to release glycogen. This is converted into glucose, thereby lifting levels of blood sugar to normal.

## **What is known about insulinomas?**

In most cases a single tumour develops in the beta cells (the insulin producing cells) within the pancreas. In a very small number of cases (1-3%) the tumour will develop in insulin producing cells that have 'escaped' into the abdomen. The reason why the tumours form is not yet fully understood. They may be present for many years prior to diagnosis as the symptoms they cause can be confused with other conditions.

## What are the key symptoms?

The tumours are associated with episodes of low blood sugar (hypoglycaemia) that can cause an array of symptoms.

Low blood sugar levels can affect the central nervous system (the brain and spinal cord) causing episodes or attacks of confusion, panic attacks and even personality change.

The insulin producing tumours can also affect the autonomic nervous system (that regulates many of the organs, muscles and systems in the body, such as heart rate) causing palpitations, sweating and a trembly feeling.

Patients with an insulinoma will find eating or drinking something sugary can quickly alleviate their symptoms.

### *Low blood sugar levels caused by the tumour can trigger*

Confusion, anxiety, personality disorders and even aggressiveness

Rapid heartbeat, sweating, palpitations, feelings of hunger, dizziness and drowsiness. You may look pale, have a headache, feel irritable and suddenly weak.

If blood sugar drops very low or very suddenly it is possible to lose consciousness and it can even lead to seizures (fits).

**Note:** Insulin normally helps to lower blood sugar, and when it gets to the right level the body gets a signal to stop the insulin production. In people with insulinomas the body does not get this signal and high levels of insulin continue to be released leading to the hypoglycaemic episode.

## Diagnosis

Insulinomas can be diagnosed through a simple fasting blood test. Your NET specialist will look for a certain combination of:

- low blood sugar (less than 2.8 mmol/l)
- high insulin (6 microunits/ml or higher)
- and high levels of C-peptide (0.2nmol/l or higher), an inactive amino acid that in a healthy body will be produced in equal amounts to insulin. I thought it was a byproduct of insulin production. I don't think it is an amino acid

The doctor may also use a 'rule of thumb' guide called the Whipple's Triad. Under this guide an insulinoma will be considered if you experience:

- Symptoms and signs of hypoglycaemia
- Blood sugar levels below 2.8 mmol/l
- Recovery from an attack after eating something sugary

If you are on medication for diabetes you can still be tested for insulinomas.

If someone takes insulin, doctors will be looking at the levels of C-peptides in the blood. Commercially used insulin does not contain C-peptides, so a test will look for certain levels that might suggest a tumour is present.

If someone takes sulphonylurea tablets (that lower blood glucose levels) the doctor will be looking at the level of sulphonylurea in the blood in relation to insulin, blood sugar and C-peptide levels. If it is normal an insulinoma will be suspected.



## The long fast

If further confirmation is needed you may be invited into hospital for a special fasting test that can take between 48 and 72 hours. You will not be allowed to eat or drink, apart from water, throughout this period. You will have blood tests at intervals of between three to six hours, and also whenever you show symptoms of low blood sugar, to look at the key levels of blood glucose, insulin, C-peptides and sulphonylurea. This hospital fasting test will diagnose insulinomas in more than 90% of cases.

Low blood sugar levels caused by the tumour can trigger  
Confusion, anxiety, personality disorders and even aggressiveness  
Rapid heartbeat, sweating, palpitations, feelings of hunger, dizziness and drowsiness.  
You may look pale, have a headache, feel irritable and suddenly weak.

If blood sugar drops very low or very suddenly it is possible to lose consciousness and it can even lead to seizures (fits).

Note: Insulin normally helps to lower blood sugar, and when it gets to the right level the body gets a signal to stop the insulin production. In people with insulinomas, the body does not get this signal and high levels of insulin continue to be released leading to the hypoglycaemic episode.

## Further tests

**SRS Somatostatin Receptor Scintigraphy (Octreotide scan)** – this is a common scan for NET patients. It can help to detect tumours that might be missed on other conventional scans. Around 50% of insulinomas have special receptors on their surfaces called somatostatin receptors.

Octreotide is a somatostatin analogue, a substance that mimics the action of naturally occurring somatostatin. When Octreotide is combined with a mildly radioactive agent and then injected via a vein in the arm, it binds to somatostatin receptors on the tumour surface and the tumours ‘light up’ on the screen as hot spots. This is a useful test to find out more about your tumours: where they are positioned, and also whether you would be suitable for certain treatments that use Octreotide or another chemical called Lanreotide as a carrier agent.

**Ga-68 octreotate/octreotide PET scan** – this is a new type of molecular imaging for NETs. It is much more sensitive than Octreotide scan, especially for small size tumours such as insulinomas. Also, it can be completed and provide very good quality images within hours, whereas an Octreotide scan needs two days to be completed. Unfortunately, this new imaging modality is not widely available in the UK yet.

**CT scan** – a computerised tomography (CT) scan provides a three dimensional picture of the inside of the body. It can be used to determine the position and size of tumours, and regular scans are useful to find out more about the rate of tumour growth and how your tumour is responding to treatment. When you arrive at the clinic you will probably be asked to drink a litre of fluid that contains a contrast agent that helps to highlight tumours, and you may also have a cannula inserted through which a special contrast dye is administered during the scan. These both help your specialists to read the scans more clearly as the tumours are highlighted.

**MRI scan** – this is a whole body scan. Magnetic resonance imaging (MRI) scans can help reveal where tumours are positioned. It uses magnetism rather than X-rays to take pictures of inside the body. Scans can take up to one hour to complete and you have to stay very still inside the scanner lying on a couch. MRI is often used in conjunction with CT and SRS scans. They have a good value when detecting liver metastases (spread of tumour into the liver).

**Endoscopic ultrasound (EUS)** – is a technique that uses a special endoscope that has an ultrasound machine at the tip. Endoscopy refers to the procedure of inserting a long flexible tube via the mouth or the rectum to see the digestive tract, whereas ultrasound uses high frequency sound waves to produce images of the organs and structures inside the body such as the liver and pancreas. Using the EUS scope, doctors can place the ultrasound probe in the gastrointestinal (GI) tract, close to the area of interest, so that a very detailed image of the deeper layers of the GI tract, surrounding lymph nodes, blood vessels, and organs can be obtained. Biopsies can also be taken at the same time. EUS is performed under sedation in the endoscopy unit. For detection of the primary tumour this procedure has a high sensitivity and accuracy level.

**Portal vein sampling** – there are other interventional tests that can be done if there is still doubt. A procedure that involves testing blood taken from the portal vein (that carries blood from the GI tract to the liver) following an injection of calcium (that stimulates insulin secretion) can reveal insulinomas smaller than 1cm. This test is called PVS (portal vein sampling) and is done under sedation.

Once the diagnosis of insulinoma has been made, it is likely that you will be screened for the presence of MEN1 (Multiple Endocrine Neoplasia, type 1) syndrome, a condition that includes tumours of parathyroid glands, pancreatic neuroendocrine tumours and pituitary tumours.

*What does screening for MEN1 involve?*

A blood test to estimate levels of calcium and certain hormones in the blood.

## Treatment for insulinomas

### Benign tumours (with no spread beyond the pancreas)

Complete surgical removal of the insulinoma from the pancreas can provide a cure. Most patients have single tumours that can be totally removed (enucleated) without even the need to cut away any part of the healthy pancreas. Often this can be performed via keyhole surgery.

During surgery the specialist may manually manipulate the pancreas and perform an intraoperative ultrasound to ensure that there are no other small tumours within it, close by or whether any lymph nodes have been affected.

### Malignant tumours

There are treatment options if you have a malignant tumour that can lead to an improved quality of life and good control of symptoms.

**Surgical** – surgery may still be considered. It may be possible to resect (surgically remove) part of the pancreas containing the tumour and also surgically remove any tumour which may have spread to the liver, which tends to be the main secondary site for these tumours.

**Medical management** – if surgery is not possible you may be prescribed a tablet called Diazoxide that can help to elevate and control blood sugar levels.

### Other treatments

**Radionuclide targeted therapy** – also known as magic bullet treatment, is considered for patients who have advanced inoperable tumours that have positive uptake on the Octreotide scan. This treatment carries a radioactive particle e.g. Yttrium-90 or Lutetium-177 attached to Octreotide to wherever there are tumour cells (which have lit up on the Octreotide scan).

**Transarterial chemoembolisation** – may be considered if there has been spread of disease to the liver. This procedure involves cutting off the blood supply to the tumours with or without the addition of intra-arterial chemotherapy. Occasionally other ablation techniques such as radiofrequency ablation might be used if the tumours in the liver are small and few in number. This involves guiding a special needle electrode to the tumour and then passing a radio frequency current through it to heat the tumour tissue and ablate, or eliminate, it.

**Chemotherapy** – can be helpful for highly aggressive tumours that do not respond to other measures. Your NET specialist will advise you. The combination usually used is 5-fluorouracil plus cisplatin and streptozotocin.

**New molecular treatments – Everolimus.** Everolimus is a new oral agent, which belongs to the group of drugs that inhibit the ability of the neuroendocrine tumour to produce new vessels. There is now evidence that the drug is beneficial in patients with pancreatic neuroendocrine tumours and, according to two small published series of patients, it can improve the blood sugar levels, if other treatments have failed to do so. However, as this treatment may have some side effects, your NET specialist will advise you whether you are suitable for this drug.

### **What about somatostatin analogue injections?**

Some NET patients are helped with regular injections of octreotide or lanreotide. But in the case of people with insulinomas this injection can make symptoms worse by decreasing blood sugar further. You will need to seek specialist advice from a NET consultant to find out whether this treatment would be suitable for you.

## **Diet for insulinoma**

Insulinomas are rare tumours, therefore there is very little research or existing evidence regarding diet in this condition. Although diet cannot control the release of insulin from the tumour, it can help prevent low blood sugars. People with insulinomas tend to suffer from low blood sugar levels until the tumour has been treated to stop it releasing excess insulin. If you are losing weight, please ask to see a dietician.

## **Glycaemic index**

The glycaemic index (GI) is a measure of how quickly foods that contain carbohydrates will raise blood sugar levels. Foods are given a GI number or classed as low, medium or high GI.

Low GI carbohydrates are released slowly into the blood and therefore are able to maintain blood glucose levels for longer. It is recommended that you try to choose low GI carbohydrates as much as possible during the day to prevent sharp peaks and troughs in your blood sugars.

High GI carbohydrates are released very quickly into the blood and are very useful when you are experiencing a hypoglycaemic (very low blood sugar) episode.

Eat breakfast cereals based on oats, bran and wholegrain wheat e.g. All Bran, Bran Flakes, porridge.

Use breads with wholegrain, stone ground flour or sour dough e.g. multigrain, wholemeal, soya and linseed, pumpernickel

Include wholegrain pasta, noodles, pearl barley or quinoa.

Choose new potatoes, sweet potatoes or yam in preference to other potatoes, leave the skins on if you can.

Eat plenty of vegetables and salads.

Add beans, lentils and other pulses to soups, stews, salads and other dishes.

Use basmati or brown rice, rather than Thai, jasmine, sticky or short grain rice.

Choose grainy crackers and crisp breads e.g. Ryvita Seeded, oatcakes.

Tips for low GI meal choices:

### **Breakfast**

Muesli\*, All Bran, Sultana Bran, Special K

Oat-based breakfast cereal and fruit

### **Lunch**

Baked beans with jacket potato

Lentil-based soup

Variety of wholegrain breads: pitta bread, breads made with mixed grains and pumpernickel

Grilled chicken, salad, basmati rice and peas

#### Evening meal

Brown rice, sweet potato, buckwheat, bulgar wheat, pearl barley, noodles

Vegetables with meal

Wholegrain pasta based meals

Beans and pulses (e.g. dahl)

#### Snacks

Fruit

Yoghurt (low fat)

Popcorn

Rye bread, fruit loaf\*

Nuts and seeds\*

\* These foods can have a higher fat content, therefore consume in moderation if you are trying to lose weight.

## **Tips on how to manage hypoglycaemia\***

**Hypoglycaemia**, or hypo, is the medical term for low blood glucose levels – that is a blood glucose level of less than 4 mmol/l. This is too low to provide enough energy for your body's activities. Hypos are one of the most common symptoms of having an insulinoma. It is caused because the insulinoma produces unregulated insulin.

What causes a hypo? A hypo can be caused by the following:

- too much insulin
- a delayed or missed meal or snack
- not enough food containing carbohydrate
- unplanned or strenuous activity
- drinking too much alcohol or alcohol without food
- sometimes there is no obvious cause

Signs of a mild hypo

Most people have some warning signs when their blood glucose level starts to go low. These include:

- feeling hungry
- trembling or shakiness
- sweating
- anxiety or irritability
- going pale (greyish complexion if you are Black)
- fast pulse or palpitations
- tingling of the lips
- blurred vision

Signs of a more severe hypo

- difficulty in concentrating
- vagueness or confusion
- irrational behaviour

### **Immediate treatment**

Once you notice your hypo warnings, take action quickly or it is likely to become more severe, and you may become unconscious or have a fit. Immediately treat with a 10-20g of a short acting carbohydrate such as:

- a glass of Lucozade or non diet drink



- three or more glucose tablets
- five sweets, e.g. jelly babies
- a glass of fruit juice

The exact quantity will vary from person to person.

If your hypo is more severe and you cannot treat it yourself, someone else can help you by: applying GlucoGel (or treacle, jam or honey) on the inside of your cheeks and gently massaging the outside of your cheeks.

If you are unconscious, Glucagon can be injected if the person you are with has been trained to use it. Otherwise the people you are with should call an ambulance immediately.

**Important:** If you are unable to swallow or unconscious, you should not be given anything by mouth (including GlucoGel, treacle, jam or honey). Make sure your family and friends are aware of this. If you are unconscious, you should be placed in the recovery position (on your side with your head tilted back) so that your tongue does not block your throat.

### **Follow on treatment**

To prevent your blood glucose levels dropping again, you should follow your sugary foods with 10-20g of a longer acting carbohydrate such as:

- half a sandwich
- fruit
- a small bowl of cereal
- biscuits and milk
- the next meal if due

The exact quantity will vary from person to person.

### **Hypos at night**

If you are concerned about nighttime hypos, check your blood glucose between 2am and 3am when hypos are most likely to happen. Keep something sugary by your bed just in case. Alternatively have a snack before bedtime such as biscuits and milk, half a sandwich, fruit or yoghurt. If your blood glucose is low before going to bed ensure that you eat to raise your levels.

### **Hypos and physical activity**

Physical activity lowers your blood glucose level so it is important to eat some form of carbohydrate before, possibly during and after your activity especially if it is strenuous or lasts a

long time. Hypos can happen up to 36 hours after strenuous or prolonged physical activity so you might need to adjust your medication or carbohydrate intake to compensate.

### **Hypos and driving**

Always test your blood glucose levels before driving. If you feel you may be going hypo, pull over, stop the car as soon as it is safe to do so and remove the keys from the ignition. Leave the driving seat and treat your hypo in the usual way. Do not attempt to start driving again until you are sure your blood glucose levels have risen again. Should they be driving if having hypos? Suggest they inform DVLA in UK?

### **Hypos and alcohol**

Drinking a lot of alcohol or drinking on an empty stomach makes a hypo more likely to occur. The outward signs of a hypo are also very similar to those of being drunk. Always have something to eat if you are drinking alcohol, and tell the people you are with about your diabetes and what to do if you need help treating a hypo.

### **Important points to remember:**

Always have something sugary with you for use in an emergency.

Wear some medical I.D.

Tell your friends or family what signs you have when you go hypo and how to treat it, as you may not be able to think clearly when your blood glucose goes low.

You will come to recognise your own hypo warning signs, but these may change over time, so be prepared to check your blood glucose level if you experience any unusual symptoms.

\*This information is from 9825/0110/a ©Diabetes UK 2010 it was produced in relation to hypos for diabetics. It has been reproduced here as it is perhaps the best advice currently available for managing and treating hypoglycaemia for those with an insulinoma.

## **Multidisciplinary teams**

Insulinoma care can be complex, and for the patient the journey can encompass not only a whole host of emotions, but also a whole range of investigations, treatments and healthcare professionals. The very fact that there is often not just one treatment option at diagnosis and throughout the patient journey, means that there has to be a collaboration among all key healthcare professional groups, who are making clinical decisions for individual patients.

This collaboration has been termed an MDT (multidisciplinary team). This is a formula that is now being used across the world in the care of cancer patients.

A patient may see some or all of the following people:

- Oncologist
- Surgeon
- Endocrinologist
- Nuclear Medicine Physician
- Radiology staff
- Dietician
- Nurse Specialist
- Palliative Care Team
- Pain Team
- General Practitioner/Practice Nurse
- Counselling Staff
- Various Technicians
- Clinic Staff
- Hospital Staff
- Hospice Team

Patients can feel more confident in the knowledge that all aspects of their care have been discussed and that the best possible treatment plan will be formulated. A well coordinated and disciplined MDT is a very important aspect for care when striving to achieve the best quality of life and the best outcome for insulinoma patients.

## Clinical Research

Research is a step-by-step process that involves collecting and examining information. Research into insulinomas is vital to improve our understanding of the disease and how it can be treated.

Research goals include:

- Understanding what causes insulinomas
- Understanding how insulinomas form
- Formulating more effective diagnostic scans and tests
- Discovering new treatment options, and ensuring that current treatments are being implemented to provide the best therapeutic benefit

Insulinomas are a rare form of cancer, and there are small teams of dedicated medical professionals around the world who treat patients every day. It is important that these specialists are allocated the resources to carry out research within their units, so that our understanding of this disease and how to treat it continues to grow.

In clinical trials, patients agree to try new therapies (under careful supervision) in order to help doctors identify the best treatments with the fewest side effects.

If patients want to take part in a clinical trial, they should discuss this with their specialist, who will know whether they are eligible.

All studies are run on strict inclusion and exclusion criteria for the safety of the patients. It can be frustrating for patients to discover that they are ineligible, but no medical professional is able to influence any decisions based on these criteria.

No one should ever include a patient in a clinical trial without his or her knowledge. A doctor, nurse or other researcher will ask for permission, and they cannot enter a patient into the trial unless that patient has given his or her consent.

To help patients decide whether they want to take part, the researchers should tell them all about the study:

- What it is trying to find out
- How they will be treated
- What they will have to do

Even after consent has been given, a patient may leave the trial without giving a reason at any time. If a patient is having a new treatment as part of a trial and then leaves the trial, he or she may not be able to continue having the new treatment. In this situation, patients would be given the appropriate standard treatment for their type of cancer.

You can find out more about current trials at:

### **Current Controlled Trials**

This website allows users to search, register and share information about randomised controlled trials. Covers multiple registers, including England, Scotland and the US.

**<http://www.controlled-trials.com/>**

### **ClinicalTrials.gov**

This is a register of federally and privately supported clinical trials conducted in the United States and around the world.

### **clinicaltrials.gov**

You can also search for research trials available to patients in the UK and Europe on these websites:

**[www.macmillan.org.uk](http://www.macmillan.org.uk)**

**[www.cancerhelp.org.uk](http://www.cancerhelp.org.uk)**

**[www.ctu.mrc.ac.uk](http://www.ctu.mrc.ac.uk)** (The Medical Research Council Clinical Trials Unit)

**[www.ncrn.org.uk](http://www.ncrn.org.uk)** (National Cancer Research Network)

**[www.eortc.be/](http://www.eortc.be/)** (European Organisation for Research and Treatment of Cancer)

## Part 3

### Finding help and support

[www.insulinoma.co.uk](http://www.insulinoma.co.uk) is a website created to support all of those diagnosed with an insulinoma. The overall aim of the site is to establish an insulinoma community and to provide up to date, easily accessible information. The website provides a unique platform by bringing together information and people who have an insulinoma. The more we share the more we learn.

#### Join us on Facebook

The Facebook insulinoma page is an excellent and current way to ask any questions that you may have about insulinomas as well as to meet others with an insulinoma.

To access the Insulinoma Facebook Group- copy this link into your browser window:

<http://www.facebook.com/groups/15631450139/>

#### NET Patient Foundation

The NET Patient Foundation continues to be the only charity in the UK and Ireland that offers information and advice to those affected by neuroendocrine tumours, which includes expertise about insulinomas.

Patient support and advice: 0800 434 6476

<http://www.netpatientfoundation.org>

#### AMEND (Association for Multiple Endocrine Neoplasia Disorders)

Support and information for people affected by Multiple Endocrine Neoplasia Disorders and associated endocrine tumours and syndromes

Tel: 01892 516076

<http://www.amend.org.uk>

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