Being Cortisol Dependant.

By Benedict Sloane

The first thing to get my head around was that I would have to take hormone replacements for the rest of my life. This alone was a lot to take in. I initially thought I would just have to take them for a short while, like a course of antibiotics, but I soon learnt that this was forever! At the time swallowing tablets was difficult for me. I remember being given vitamins and cod liver oil tablets as a child and had such great difficulty taking them, that I pretended to take them and then dispose of them. I soon got used to taking these though; I had to, as they are essential for my survival.

Cortisol, also known as Corticotropin or ACTH, is the body's natural steroid hormone. It is the body's stress hormone and is vital for life. Although all the hormones are essential for life, Cortisol plays the most complex role in regulating bodily functions and a wide range of vital processes throughout the body, including controlling blood pressure, heart rate and blood circulation and the immune system response; how your body is able to cope in times of stress, whether it be physical stresses such as an accident, viruses, bacteria and other threats, but also emotional stresses.

Produced by the adrenal cortex, (the two adrenals sit on top of each kidney), cortisol serves as a chemical messenger, so called because the Pituitary gland, often referred to as the Master Gland, sends a message to the adrenals to produce cortisol. These messengers are like pressing a button. But because my pituitary was destroyed, (there was no sign of the pituitary stalk in post-surgery MRI scans) my adrenal cortex isn't getting the messenger from the pituitary: there is no connection. As a result I have a condition called Pan-hypopituitarism. (Hypopituitarism is a deficiency of one or multiple hormones your pituitary gland makes. Pan-hypopituitarism is a deficiency of **all** of the hormones your pituitary makes, and this is what I have.) I will discuss the other hormones later, but in regards to my body being unable to make cortisol, this means I have another condition; I am completely Cortisol Dependant (Steroid Dependant) which means, that in order to survive I have to take replacement cortisol in the form of a tablet called, Hydrocortisone. I am dependant on it; without it, my body would fall into shock within hours and I would die.

Cortisol replacement is incredibly complex and cannot mimic the body's usual cortisol production. It's a fine balancing act; too little, a deficiency, of hydrocortisone, and the body cannot function and would lead to death. Too much, even a small amount, risks a host of side-effects. Normally the body regulates cortisol minute by minute, second by second. Both the hypothalamus and pituitary can sense whether the blood has the appropriate amount of cortisol circulating. If there's too much or too little cortisol, these glands respectively change the amount that gets released into the bloodstream. This is what is referred to as a negative feedback loop. Naturally cortisol production in the body begins to build at around 3am, and peaks at around 7-9am, giving us that get up and go when we wake up. There is then a further mini-peak around midday and by evening time, levels have tailed off. This is known as the circadian rhythm. Unfortunately conventional hydrocortisone replacement cannot reproduce this, so it's easy to inadvertently under or over-replace, and could at least in part explain why patients with adrenal insufficiency suffer from a poor health-related quality of life, with an increased risk of mortality, sleep disturbances, impaired psychological well being and also, at even slightly too high doses, significant weight gain and obesity, worsening of cardiovascular risk factors and defects in bone turnover. It's hard to replicate mother nature and my replacement is crude in comparison. I used to take Hydrocortisone three times a day, on waking at around 8.30am, at lunch time and in the early evening, producing three unnatural peaks of the hormone in my system, rather than the normal steady secretion. Each peak can result in further weight gain.

After some years of living with the condition I attended meetings by a Professor Hindmarsh who is in favour of splitting up the dose of Hydrocortisone further, in an attempt to mimic the body's normal circadian rhythm more closely. As a result I started breaking my cortisol into six doses throughout the day. The usual (replacement) dose of cortisol/hydrocortisone is 20-30mg daily. I currently take 22.5mg daily as follows;

- 2.5 mg at 4/5am, when I usually get up in the night to go to the toilet. This was in attempt to try and mimic mother nature. Unfortunately it hasn't given me the get up and go I had hoped, presumably because of my hypothalamic fatigue, which I will cover in the next chapter. However, one positive is that it means my body has some cortisol coverage in the bloodstream in these early hours, without which, my levels could dip dangerously low and prove dangerous, especially if I were unwell or had an impending illness or infection.

- I then take 10mg on waking at approximately 8.30am,

- and then I take 2.5mg at midday, 2pm, 4pm, and 6pm.

Personally, I haven't noticed any improvement in this new regime, including with my energy or weight, presumably because, apart from anything else, it still has nothing on the minute by minute secretion that naturally occurs. Attempting to mimic the body's usual cortisol production is challenging to get right, and as a result it's easy to inadvertently under or over-replace, and more often than not, caution is taken in slight over-replacement to avoid the risks of adrenal crisis' that come with under-replacement. This is where regular blood tests including 24 hour cortisol profiles would be useful. For a few years after my operation I had these in hospital; my bloods were taken once an hour for the whole day to monitor my levels of cortisol and then my dose adjusted accordingly. Unfortunately though, due to NHS cuts, this no longer happens. Instead, one blood test is taken 4 hours after taking my dose of cortisol on waking, which is not nearly thorough as a 24 hour profile.

It's worth mentioning at this point that there are now slow-release versions of Hydrocortisone replacement called, Prednisolone and Plenadren. Some patients swear by them, however I did not get on with them. They caused me to gain even more weight and I felt that the dose tailed off too quickly and had to top up with hydrocortisone anyway.

Despite being far from ideal, my replacement cortisol quite literally keeps me alive on an ordinary day to day basis. However, cortisol has a very important role in helping the body respond to and cope with stress, be it illness, infection, accident, surgery, or emotional stress, hence why it is also known as the stress hormone. During stress, our cortisol levels are drained to a dangerously low state, where ordinarily, our pituitary will go, hang on, we have a stressful situation here, and it sends the signal to the adrenals to produce the extra cortisol it needs to help the body respond and cope appropriately to the situation in hand. But my body cannot carry out this lifesaving defence mechanism, and this is where the danger and complexity of cortisol dependency lies. In periods of stress on the body, I will be in a very vulnerable position and at risk of a life threatening condition and medical emergency called, Adrenal Crisis, so it is critical to try and mimic the natural response to these situations of stress by increasing the dose of Hydrocortisone appropriately and immediately, because if immediate action isn't taken, deficiency of cortisol risks shock to the body resulting in an adrenal crisis, low blood pressure, suffering organ damage, losing consciousness, coma and death.

Therefore there are what are known as sick day rules, meaning how much extra replacement cortisol the body needs to cope with different various stressful situations, to attempt avoid being plunged into an adrenal crisis. With illness and infection, this is not just a one off dose, but continuously until it has completely cleared. This usually means a double oral dose, but in the case of COVID for example this is (much) higher; 80mg daily (20mg, four times daily, every six hours.)

However, ordinarily the aforementioned defence mechanism may well start occurring in the body before the first obvious signs of anything untoward present themselves, so even increasing my replacement cortisol at the first sign of anything untoward, my body will have already been drained of cortisol...Imagine a sieve as the secretion of my usual replacement cortisol dose, well if my body is under stress, this is like the sieve developing a larger hole and being being drained of cortisol quicker than usual, so the first time I'm aware that I'm unwell, and start increasing my dose of cortisol, I'm already running on empty and still ended up in crisis.

Even if I was aware that I were ill before symptoms displayed themselves, and took extra cortisol straight away, I may not know i*n what way* I was ill. For example, when I had what actually turned out to be COVID, I was already taking a double dose of cortisol because I hadn't been feeling well. I took a COVID test which proved negative, so I didn't increase my dose further. The double dose would have been sufficient to cope with the stresses of a cold for example, but wasn't enough for my body to cope with the demands, the stress, that COVID was having on my body.

Often, the first I've known that I'm in an adrenal crisis is when I've woken myself up in the night because my whole body has been shaking uncontrollably due to feeling cold, so much so that my teeth have been chattering, even when wearing extra layers and the heating on full blast or during the summer. Bizarrely, I've learnt that this has been due to me burning up due to having developed a high temperature/fever. This has often been accompanied by being in a delirious state.

In an adrenal crisis I need a much larger dose of cortisol in the form of an emergency injection to cope with the extreme stress on my body. This also gets into the blood stream faster than my tablets do. Although life-saving, taking a one off emergency injection of hydrocortisone wouldn't simply be the end of the matter; upon any circumstance where I needed this emergency injection, I would need immediate hospital admission, ensuring continuous intravenous cortisol replacement at the appropriate higher dosage, in line with sick day rules, so my body could cope with the ongoing demands of the illness; the stress on my body. This would need to continue alongside hydration and close and constant monitoring until I'm completely stabilized, definitely through the crisis and certain I've fully recovered. Only at this point should I be discharged and my dose be reduced, and this must never be done suddenly, but tapered down in a gradual way so my body can adjust and not risk a set-back. On a recent admission with adrenal crisis I was discharged too quickly, only to dip into a crisis again that evening, and I needed to be admitted once again.

It's a fine balancing act when it comes to reducing my dose following illness because reducing the Hydrocortisone too rapidly risks sending me back into crisis. Whilst higher doses of hydrocortisone are life-saving when needed, continuing the increased dose for any longer than necessary can be detrimental to my health, including cardiovascular risks, high blood pressure, bone loss, and significant weight gain in a relatively short period of time. This proves a particular challenge during prolonged illnesses, such as long-lasting colds or indefinite emotional stresses on the body, as I'll cover later in the chapter. I can't exactly be on a permanent double dose, but then again, how do I prevent an adrenal crisis? Following a recent adrenal crisis it was advised my dose of Hydrocortisone be kept permanently increased. As a result I gained a huge amount of weight, over 3 stone, in a matter of weeks.

My weight gain is a very sensitive and complex life-long battle for me since my operation, with many factors at play, with cortisol replacement being just one. Cortisol controls metabolism, how your body breaks down carbohydrates, proteins and fats and helps to control the blood sugar level to give you more energy. In a future chapter I'll cover the other weight gain factors at play, mostly which are a result of damage to the Hypothalamus; Hyperphagia and hypothalamic obesity. Additionally, a secondary factor on my weight is hypothalamic fatigue which hugely limits my capability to exercise and being active, which is a key part of weight management. I covered this fatigue extensively in the following chapter.

I always carry the emergency injection on my person at all times, and have a spare at home. It is a real life-line, and as with all my hormone replacements, its like an extension of me. It is also important to stress that if there is any doubt, administering an emergency injection won't cause any harm, but without it when needed, my body would fall into shock and I wouldn't be able to survive. If I were unconscious, shaking or delirious, or if there was some other reason I'd be unable to administer it myself, or have no advocate with me, then I would have to rely on a paramedic or someone else to do so. I always wear a medic alert necklace and carry a steroid emergency card on my person, and although it's in the paramedics' training to check for them, there's always an additional a fear lingering at the back of my mind though that it could be missed. Additionally, having to wait for someone to administer it could set me back life-saving minutes.

Sometimes there are circumstances, such as headaches, common coughs & colds, and some emotional stresses that the sick day rules say increased cortisol isn't required, however I find that it does, as else I feel so unwell that I'd risk a crisis. I find when I get a headache for example, a typical symptom of Pan hypopituitarism, of which I get frequently, only a nights sleep, along doubling my cortisol will relieve it. Having lived with this condition for so long now, I've become my own expert on my condition so I know when my body needs extra cortisol. This brings me to a reoccurring theme in my writing, that everyone with my condition is different.

Even a common cold can easily last a few months long with my condition, It's no wonder then that I can barely recall a time when I haven't been unwell. It can often feel like I've got a constant cold. Indeed I'm considered vulnerable; being cortisol dependant I have a compromised (weakened and lowered) immune system which makes me more susceptible to illness and infection in the first place. During a cold, I've had to be even more cautious as not to overtire myself, or I risk setting myself back further. As you will learn in the next chapter, my daily activity is already extremely limited due to fatigue. When I'm unwell, even mildly, this is diminished even further to practically nothing, just complete rest. My ability to diet, in order to manage my weight gain due to hypothalamic damage and hormone imbalance, is also compromised during these periods as I find myself needing more energy to cope with the demands that the illness is having on my body.

Sometimes I have averted crisis but it's taken a lot out of me. During a recent cold (January/February 2023) I had doubled my dose of Hydrocortisone. After some days had passed I thought I was feeling a little better so I started to gradually reduce my dose back to normal. I was out for a short walk when I experienced a lot of the other vast and varied symptoms of an adrenal crisis, or warning signs of an impending one; It's my head where I first and predominantly notice when I've over done things, that something isn't right, and I'm at imminent risk;

I suddenly became weakened, crippled, by my fatigue, quicker than usual. My foot steps felt clunky and clumsy. I start dragging my feet and then swaying and falling all over the place. I was later told I was shaky and looked pale. I noticed my hand shaking uncontrollably. My head felt like it was under-water or under pressure and like my head, my brain was drowning. My head felt tight, dizzy and a cross between heavy and light-headed. I experienced what I can only describe as an out of body sensation, as if I was becoming detached from my body; I was there, but I wasn't there. In these experiences, it feels like my life force is being sucked out from me. My head felt that it was up so high, that it was a long way from my feet. I was having difficulty speaking and communicating coherently; I was muttering gobbledygook, laughing at myself and pointing to the sky. I felt delirious, and, what I can only describe as what It might feel like to be drunk and under the influence of drugs. (This brings me onto the worry that when I'm experiencing these crisis or pre-crisis symptoms that it could be indeed be mistaken for me being drunk or on drugs, which could delay or prevent the urgent care that I'd so vitally need.) I'm so tired that I've been walking for a few seconds at a time with my eyes closed. I noticed I was rubbing my face down with my hand. I recognise this as something I do when I'm overtired, presumably as an attempt to refresh myself. My concentration and awareness has been hugely compromised resulting in additional dangers, for example, walking close to the pavements' edge and not looking properly, or at all, when crossing roads. Even when I was able to sit and rest I experienced continued clumsiness; dropping/spilling/bumping into things. At rest I also experienced another sign of over-tiredness in which I found myself staring in the abyss, and entering a trance-like day dream.

A lot of these symptoms exhibit the hallmarks of low blood sugar levels, and it is the case that in these situations that I crave something to eat and drink, almost ravenously, alongside the necessity for extra cortisol and urgent and immediate rest/sleep to avoid crisis. It was clear that I had started to decrease my dose of Hydrocortisone too soon; after an initial boost of a whole 10mg tablet, I resumed my double dose until I felt more confident to try gradually decreasing it back to normal again.

Although it was vital for me to sleep to avert an adrenal crisis, a crisis can also be dangerous, and perhaps overlooked because all I want to do is sleep. Therefore it's important I have someone with me or who can regularly check on me when I'm in this state. Whilst a couple of hours later I felt I was through the initial danger zone, I dipped in and out of these symptoms for the next week, where I felt I was constantly on the knife edge of falling into full blown crisis.

This dizziness, and other symptoms I've experienced in my head (particularly on standing), the severe weakness and the confused and delirious state are all recognised signs of an adrenal crisis. As you will read in further detail in the next chapter, I have ended up feeling unwell in this way even when I haven't had an infection, physical illness or trauma, but simply through tiredness and over-doing things; exhaustion is a symptom, but also a trigger for an adrenal crisis.

I also start to feel this way in the very rare occasions I forget to take a regular daily dose of Hydrocortisone. This of course prompts me to take it. More so, this is the case with my new regime, where, although I take the hydrocortisone more frequently, each dose is smaller, and therefore its lifespan, and what is known as it's 'half life' is less.

Even a sore throat can be dangerous in my cortisol dependant body. I have to be very vigilant as this has been a warning sign of something more sinister. Sometimes I've had a raging sore throat but not felt a thing, and on two occasions I thought I had a sore throat, but it actually turned out to be tonsillitis.

Although not as obvious a sign, dehydration is also a symptom of adrenal crisis. As I'll discuss later, I'm at additional risk of dehydration due to another condition, Argine Vasopressin Deficiency, as a result of being in deficient in another hormone, Desmopressin/DDAVP, which effects water balance.

Vomiting is also a sign of adrenal crisis, but also a danger sign even if I'm not in crisis because if I can't keep my cortisol down to be absorbed, then I'd need it in the form of the emergency cortisol injection to avoid a crisis. If I was in an accident or emergency, even if I was conscious, I would need the emergency injection of Hydrocortisone administered urgently. This would be critical for me to cope and hopefully avert my body going into shock. Bear In mind, in such a situation this extra cortisol would normally be produced automatically and immediately, so the time it takes me, or someone else, to get the injection set up, injecting it, and for it to get into my system, all whilst in a situation likely to be filled with panic, will have already set me back crucial minutes.

There are so many situations where the body would normally produce extra cortisol, for example during operations/surgery, even dental surgery, I would need extra cortisol before, during and afterwards for my body to cope with the added stress on the body. The amount would vary depending on the nature and seriousness of the operation.

Shocks like someone coming up behind me to surprise me, or someone running past me aren't good for me either. I can't cope even playful play-fighting. I feel fragile.

In the event of any accident, however small, I would need extra cortisol to cope and prevent my body falling into shock and risking my life. Here's an example;

(February, 2021) I slipped and fell on some ice. I ended up on the floor, where I stayed for a few minutes before I was able to get up. I didn't suffer any physical harm, other than a barely noticeable graze to my hand and knee, but I noticed almost instantaneously that I was in shock; feeling shaken and slightly out of body. I was aware that my body wouldn't be able to produce the extra cortisol to compensate for the situation that had just occurred, and therefore would need extra Hydrocortisone immediately. Despite taking this straight away I felt shaken and out of body for a few hours. Whilst I averted hospital admission on this occasion, I had to completely rest and I couldn't do a thing for the next 48 hours, throughout which time I had to double my dose of Hydrocortisone. This is just to provide an insight into how a relatively small physical accident/stressful situation, where anyone else would have just got up, brushed themselves down and carried on, can affect my cortisol deficient body.

As well as physical stress, its important not to underestimate the demands and impact that emotional stress has on the body. Emotional stress can precipitate symptoms of cortisol deficiency and the need for stress doses. It's worth mentioning that 'good' stress, including celebrations such as Christmas, is still stress and so it brings physical demands. (Addisons Disease UK Instagram page.) And this is during celebrations! So imagine when things get *really* stressful! Unfortunately with my multiple health conditions, come multiple and daily worries, fears and anxieties. We all worry of course, that's human nature, but my brain damage affected the cognitive side of things where I simply can't switch off; one thought leads to another, overanalysis and worry. Thoughts and fears constantly plague my mind. It's a constant battle, a balancing act to try and overcome this though, as I can literally make myself seriously physically ill through emotional worry and anxiety. When i get stressed I can almost feel myself lacking the extra hydrocortisone that I should be producing, desperately seeking stores that just aren't there, risking the aforementioned symptoms.

My health suffered further when I endured a benefit ordeal that lasted for over a year! This resulted in hospital admission with adrenal crisis due to Tonsillitis, brought on through the unimaginable amount of extreme and prolonged stress I endured. This also didn't help my existing PTSD. It left me traumatised further, and in one sense the ordeal hasn't ended...I'm at a constant and heightened stress about when, not if, I'll next hear about my benefits again...I fear the postman and the mail coming through the door. This uncertainty and heightened anxiety isn't good for anyone's health, but more so being cortisol dependent. More stress comes with the governments increased hostility to those with disabilities and their proposed changes to the benefit system to make it even more difficult to prove our disability and our incapability to work including re-writing the rule book altogether. I often say my disability and fighting for the benefits I'm entitled to are like fitting a square peg through a round hole, only the size and shape are constantly changing. It's utterly frustrating and exhausting, and creates a constant anxious state, and putting me at risk of adrenal crisis. I've sometimes only just managed to temporarily control my stress around all this, and all it takes is the mention of the word 'benefit', or a related word, to set off a trigger of falling dominoes of worries in my mind. My whole days can become consumed by it, so much so I find myself at risk of another danger in not being fully aware of my surroundings. It consumes me. It's never-ending. It's no way to live.

Adrenal Insufficiency is covered by the Equality Act 2010's definition of disability. This is because adrenal insufficiency is a life-long condition which can seriously affect a person's ability to do normal day-to-day activities and is fatal if left untreated. (Addisons Disease UK Instagram page) You'll discover just how limited my day to day activity is in the following chapter regarding Hypothalamic fatigue.

As you can see it is all very complex living in a cortisol dependant body. But the talk of adrenal crisis and hospital admission brings up a genuine everyday concern in living with my condition; that it is just not understood, even by many medical professionals. If I were to become ill and needing admission to hospital for treatment, what would happen to me? I have an amazing pharmacist who understands and, in many ways has been on this journey with me. But even in a hospital setting, perhaps even more so, I do not feel safe. I have to say, the Paramedics have always been amazing, but once I'm transferred to hospital it's another matter. Adrenal insufficiency and cortisol dependency can be easily overlooked and this is a constant fear of mine. An added complication is that some people are cortisol deficient, meaning that they might still produce some cortisol themselves, where as I don't produce any! This means I am ***completely*** **cortisol dependant**!

One issue is that my disability is hidden so I look 'normal' on the outside but this is just one part of the problem...there is a distinct lack of understanding and knowledge that such a condition even exists. To put it's rarity into context, I was once told that most G.P.'s will never come across a case of cortisol dependency in their whole career. Indeed mine had not before my case.

Even if one nurse or doctor has it explained to them, and yes they often need this explained, or in rare occasions where it is understood, this never seems to be passed on as I'm handed over through the system into different departments and eventually a ward. This breakdown of communication is deeply worrying and a constant fear. So far my Mother has been an incredible advocate for me, bringing my Pan Hypopituitarism, particularly my cortisol dependency, to the attention of hospital staff; doctors and nurses. But she can't and won't always be able to, and she shouldn't have to! She even put herself at risk when I had COVID in joining me in the hospital, then catching COVID herself.

Apart from anything else, the lack of understanding about just how life dependant my cortisol replacement really is means that it is often not administered on time. This was demonstrated during one of my last adrenal crisis' where there was poor transition of care when I was transferred from one hospital to another. I knew that I would be due my next intravenous injection of hydrocortisone shortly after being transferred. My Mother was not with me because upon transfer she had to drive home and get some much needed rest as we had been at the previous hospital all day long. Whilst I will not always be able to rely on her, it highlights a valid concern; that without someone acting as an advocate, someone to speak for me if I wasn't able to, what would happen to me? In a crisis state I could be unconscious, or if conscious, unable to speak clearly or coherently. I fear that even if I'm coherent, I will still not be heard, because there is an air that the medical professionals know best and don't always like things being brought to their attention. I genuinely fear that my life-saving treatment will be missed. This is a constant worry...

...The time for my next dose of cortisol came and went and I became increasingly concerned. The paramedics who transferred me were as great as always but when they handed over care, I was just lost, hanging in limbo. I was supposed to be transferred into the continuing care of endocrinology at the hospital I was transferred to, who I was told, were expecting me and would be ready and waiting but I ended back in triage, effectively starting all over again in a general waiting room where I knew I was hours from being seen, had I not tried to speak to someone. I tried to talk to a couple of doctors behind a counter and I explained the situation and that my IV cortisol was, at first due, then overdue. My explanation presented glazed expressions. I tried to explain that this was an emergency as this hormone, cortisol, is vital for life, but still nothing! Over an hour after my cortisol was due I had become increasingly worried. Eventually my repeated appeals for help were responded to by a female doctor behind the counter who laughed and told me I was 'over-reacting'. This is the response when I was clearly and rationally stating my the facts and my concerns, albeit understandably in an increasingly anxious state! Although i knew it wouldn't be nearly a high enough dose, I was seriously contemplating taking some of my oral cortisol when I was *finally* seen and given my IV cortisol just shy of two hours after it was due, by which time I was feeling incredibly unwell and dipping back into the symptoms of a crisis that I was trying to overcome and the reason for my hospital admission in the first place. I was seriously becoming concerned for my life and with no-one to turn to who understood the severity of the situation, I wouldn't know what I would have done. What if I wasn't coherent, or conscious to be able to bring this to someone's attention, would I still be here? And even if I was, like in this situation, would they even listen?

At pituitary meetings some of the best doctors in the field have admitted that despite all their knowledge, they aren't the ones living with the condition, therefore they will never know what it is like to live with it. As I've learnt, I'm my own best expert on my condition. I have to advocate for myself even when I'm really too ill to, when I *should* be confident that I'm in safe hands. I think the one thing that really struck me by that incident was that even some health professionals don’t realise that complete cortisol dependency exists, and the real danger this poses.

Fortunately The Pituitary Foundation have run campaigns aimed at healthcare professionals to try and increase the awareness of Adrenal Insufficiency, and Hydrocortisone as life-sustaining replacement for patients with the condition. There was a campaign aimed at Paramedics and my experience with them has been widely positive, but I feel a similar campaign for A&E staff is desperately needed to provide real change and peace in mind for people like myself.

There are so many rare conditions. Mine is just one. I believe the reason why this happened to me, my purpose in life, is to create awareness, so my condition is more understood, so those living with rare and hidden disabilities can live their lives without hostility and fear, that patients like me don't risk losing their lives. If my rare condition becomes just slightly less rare by more people being aware of it’s existence and seriousness then I feel my life has been an accomplishment, more than I could ever have dreamt.