



Submission to the Royal Commission into Violence, Abuse, Neglect and Exploitation of People with Disability

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Australian Addison's Disease Association Inc. (AADA)

Raising Awareness of Addison's Disease & Supplying a Caring Network for Members and their Families

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Executive summary

As with many rare diseases, the key stages for many people with Addison's disease and adrenal insufficiency (AD/AI) include periods of being undiagnosed, diagnosis, treatment, and management. Many people live normal lives with AD/AI once they are diagnosed and adequately medicated. Others struggle on a daily basis, and this can be even more problematic when people have comorbidities. Often people experience temporary disability prior to diagnosis, and some people have horrific diagnosis stories.

As an Association, we hope that contributing our stories will raise the Commission's awareness of temporary and permanent periods of disability that can arise due to being misunderstood or neglected or unreasonable assumptions made by the medical profession.

The Committee invited the AADAI members to share their stories of 'neglect' they felt they experience in their AD/AI journey (see Attachment 1 for the invitation to members). Seventeen stories were shared by members. The Committee has provided a synthesis of the common *themes* incorporating excerpts of the member stories and *recommendations*.

Themes:



Neglect due to insufficient doctor knowledge



Neglect during emergency treatment



Neglect during hospital treatment



Neglect during monitoring and ongoing treatment



Neglect during the diagnosis odyssey



Reporting neglectful treatment

Recommendations:



APPROACH

National approach across health jurisdictions to AD/AI emergency management.



EDUCATION

Educate medical and allied health professionals on AD/AI awareness, diagnosis and treatment.



SHOWCASE

Showcase best practice for people with AD/AI to raise awareness, empathy, & knowledge.



COLLABORATE

Support collaboration between medical professionals and member associations.

Rare diseases can be difficult to diagnose as the front-line medical personnel will most likely not have seen a case before. Recognition of a cluster of symptoms beyond a medical practitioner's normal range of experience is actually an invitation to refer to a diagnostic specialist. This submission discusses one of the more common rare disease conditions so deficiencies in treatment here will point to deficiencies in the approach to all rare diseases. However, Addison's disease and secondary adrenal insufficiency are common enough that in any larger town or suburb, there will be enough cases requiring management that emergency and hospital medical staff should be on the alert at the very least for acute adrenal insufficiency or adrenal crises. Even though new cases are indeed rare, adrenal crises are frequently the route to finally being diagnosed. Because an adrenal crisis is life threatening, prompt recognition and treatment is vital.

Association members are often reluctant to admit they have a disability. However, as individuals without functioning adrenal glands, they fall within the definition of "disability is an impairment of body function or structure". The reluctance springs from a determination to live as close as possible to the functions of their peers using appropriate monitoring and medication. This submission discusses cases where that ideal was not met through neglect in diagnosis, treatment or support, particularly in emergency situations. It attempts to point a way forward through improvements in training of medical and allied health professions that may benefit much rarer conditions as well.

Almost 1 in 5 Australians live with one rare condition or another, so improvements in their quality of life and opportunity to participate as citizens will benefit us all.

Please note that the Member Stories have been anonymised to remove names of the members, hospitals and medical practitioners. Specific details are available upon request. Annex 1 is a letter from an identified hospital and is submitted separately.

1. Australian Addison's Disease Association Incorporated (AADA) and its purpose

The Australian Addison's Disease Association Incorporated (AADA) is a support group comprised of member volunteers providing valuable information and resources on Addison's disease/adrenal insufficiency (AD/AI) to the medical community, patients and carers.

The Association website is www.addisons.org.au

2. Disability and Addison's Disease/Adrenal Insufficiency

2.1. Definitions

Relevant parts of the *Disability Discrimination Act* (1992):

- total or partial loss of the person's bodily or mental function
- a disorder, illness or disease that affects a person's thought processes, perception of reality, emotions or judgement, or that results in disturbed behaviour
- includes a disability that presently exists or previously existed but no longer exists

According to the social model of disability, disability is caused by the way society is organised rather than by the person's impairment:

"The social model of disability says that disability is caused by the way society is organised, rather than by a person's impairment... It looks at ways of removing barriers that restrict life choices for disabled people. When barriers are removed, disabled people can be independent and equal in society, with choice and control over their own lives" (www.unicef.org)

Neglect includes physical or emotional neglect, passive neglect or wilful deprivation. Neglect can be a single significant incident or a systemic issue that involves depriving a person with disability of the basic necessities of life such as food, drink, shelter, access, mobility, clothing, education, medical care and treatment.

2.2. Addison's Disease/Adrenal Insufficiency and disability

"In my case adrenal crisis come[s] on very quickly, mostly without warning. Within minutes I am unable to stand or walk unaided... My mental clarity is compromised and I find it difficult to follow conversations, make decisions and maintain focus... For me, Addison's Disease can be very disabling and I require long periods of care and assistance before I am able to function independently."
(Member Story 3)

Under proper treatment, living with AD/AI means being careful to prevent and manage adrenal crises that can be brought on by elevated stress levels, illness, the complicating effects of comorbidities, and emergency situations. Adrenal crises can manifest differently for individuals. It is an invisible disease; often, symptoms are not seen by an observer, and those same symptoms are constantly self-managed by the individual. Our team of medical professionals and the health system, especially the ambulance service and hospitals, feature in many of our lives. For someone living with AD/AI, disability can be temporary or permanent.



People newly diagnosed may have undergone a long period of feeling unwell and can feel unable to carry out their normal lives. For some this 'diagnosis odyssey' (see Section 3.5) can be prolonged and may lead to death; for those who do eventually get diagnosed it can take some time to adjust their lives and find 'a new normal'.

Even with adequate treatment, people living with AD/AI have a daily battle with fatigue. Many people modify their daily activities to ensure they get enough sleep and limit stressful work environments. Workplaces can be requested by people with AD/AI to make reasonable adjustments to allow for flexible work hours or a modified environment (such as a couch in the office to allow for a rest if needed). Working at home during COVID-19 has been useful for some people to enable flexibility.

For a person living with AD/AI, friends, family, employers, and the health system all have a part to play in removing barriers to living a life without restricting choices. In particular, society determines access to quality, equitable, knowledgeable, responsive and respectful healthcare.

3. Themes based on personal lived experiences of neglect

3.1. Neglect due to insufficient doctor knowledge

People living with AD/AI rely on doctors and specialists having the knowledge to be able to recognise symptoms of adrenal insufficiency and adrenal crises, undertake the right diagnostic tests, and prescribe and educate them to enable them to self-manage the disease.

Evident in a number of the stories was reference to doctors, endocrinologists and other medical staff who lack the knowledge of AD/AI and how to manage and treat it in an emergency. The lack of knowledge stems from the small amount of time dedicated to learning about AD/AI, along with other rare diseases, during medical training. Also, because it is rare, doctors may not have the experience of treating patients with adrenal insufficiency and adrenal crises.

Common across these stories of insufficient knowledge of medical staff is an unwillingness of the doctor to listen to the lived experience and knowledge of the patients (Stories 1, 4, 6, 7, 8).

Negligence occurs where medical staff are not willing to listen to a patient's knowledge of emergency treatment procedures, or disregard or downplay the symptoms the patient is feeling because they do not know what to look for (Story 1, 7) or do not appreciate that adrenal crisis is life threatening and needs immediate treatment (Story 8).

Some observations by members of insufficient knowledge by medical staff include:

- Lack of knowledge of how to diagnose and identify the symptoms of Addison's disease (see theme 3.5);
- When undergoing procedures, there is a lack of appreciation for the need to manage adrenal insufficiency with additional medication (Story 4);
- Incorrect knowledge of the symptoms of an adrenal crisis, and as a result downgrading of the severity of the situation (Story 1), for example, when a patient has the hydrocortisone emergency injection (Solu-Cortef®) and their condition has improved but the doctor may

believe that ongoing treatment is not needed with IV hydrocortisone and saline solution (Story 6); and

- Insufficient knowledge in a crisis situation leading to a misinterpretation of the specialists' advice (Story 4) or by not appreciating the life threatening nature of an adrenal crisis (Story 17). When dealing with a patient in emergency, we have observed that a doctor can seek information about Addison's disease, adrenal insufficiency, adrenal crisis, and emergency treatment protocols by: searching on Google; consulting with the hospital's or the patient's endocrinologist/GP/ specialist; or consulting the patient's emergency management plan. This research by practitioners is encouraged by the AADAI as we do appreciate that this is a rare disease and we would prefer the attending doctor had the latest information.

Examples from Member Stories:

Member Story 1:

[On this occasion of feeling symptoms of an adrenal crisis] I'll follow my endocrinologist's advice: inject hydrocortisone and call the ambulance. Ms Paramedic arrives 20 minutes later... I say my vague symptoms are gone now. Ms Paramedic follows her rules, giving me a litre of IV saline... An hour after injecting, I feel okay. Ms Paramedic reports (accurately) to Ms Tired-Nurse, 'XXX feels okay. He complained, but we didn't see any symptoms.' Ms Tired-Nurse laughs while reporting to Dr Busy, 'Bed 1 reports no symptoms, and that's what Ms Paramedic saw.' Dr Busy says something rude, then ignores me... Finally, 3 hours after I injected myself: 'My endocrinologist told me, "You don't need to look sick to go to ED If you look sick, you're probably too late and already in big trouble". Ms Tired-Nurse decides I'm lying, stupid, crazy, or all three. Doctor Baby says 'I'm discharging you.' 2 minutes later he says, 'if you conjured up some feeling that you know more about medicine than the amazing professors who have taught me full-time for the past five years, you're more than welcome to phone and talk with the off-duty nurse about it. Bye.' ... Some hours later, at home, my wife wakes me up. I feel death approaching...

Member Story 4:

Post op, I was seen by the Junior Medical Officer who blithely told me he had spoken with my endocrinologist who had instructed him to change the type of my steroids and reduce them to less than half of my maintenance dose.

When I said he must have misunderstood my specialist, because recognised protocol is always to increase not decrease steroids during and immediately post trauma or stress, he insisted he was right... Fortunately, I was conscious, aware of my conditions and medications, and able to speak up for myself. I shudder to think what the outcome would have been otherwise.

Member Story 6:

The doctor at ER gave me 100mg of Solu-Cortef® and transferred me to ICU. I asked to see the doctor there and requested that he page the endocrinologist on duty, as they are the experts in dealing with an Addisonian crisis. Again, he said there was no endocrinologist on duty and that he would contact one in the morning.



I explained that I needed another dose of 100mg of hydrocortisone, which is the recommended treatment during an Addisonian crisis (100mg every 6 hours until patient is stable). I was only given 50mg until the doctor returned after some time (unsure how long), and reported that he had managed to get in touch with the endocrinologist registrar and was instructed to give me another 50 mg dose (total of 100mg) and organised for me to be transferred to the ward as an in-patient.

As a result of delayed medical care, I became so ill that I had to remain in hospital for 4 days and spent weeks recovering at home.

Member Story 7:

[While paramedics attending my home] My husband gave this information to one of the ambulance staff, who opened the envelope and quickly flicked through the paperwork without reading it then discarded it onto the couch saying “I don’t see a list of your medication here” ... During the time that they were at my house, I told them four times that I had Addison’s disease as did my husband on several occasions. Neither of the ambulance staff asked me any questions relating to my Addison’s disease at any time.

Member Story 17:

So I waited another 2 hours and endured 2 more ECG’s and more blood being taken which made my IV access stop working and I just resigned myself to the fact I was going to end up in ICU again as I couldn’t get the message across that this was serious and I was too exhausted and basically gave up.

The doctor eventually turned up and tried to placate me with statements like she’s in charge and there’s a lot of sick people here (my brain says and what I’m not sick???), my numbers aren’t that bad (again little voice in my head says no thanks to you that’s only because I have a pump and basically kept my system going).

Then I got the clincher, the statement that told me she really didn’t understand Addison’s disease/Adrenal Insufficiency at all, she said the nurses kept calling me but I didn’t think that you needed hydrocortisone or pain killers because you were doing so well when we sent you to short stay, I thought you would have a nap and then go home at 6am (again brain says ahh hello that’s because I had been given hydrocortisone and pain killers and they were working - thank goodness I’m the only one that can hear the little voice in my head because by this point I was totally frustrated and just fed up)

3.2. Neglect during emergency treatment

The need for emergency treatment is the reality for many people living with AD/AI. Emergency treatment is provided by general practice clinics, the ambulance service and 000 call centre, and hospitals through their emergency departments. People living with AD/AI are usually prescribed Solu-Cortef® as an injection to self-manage crises, but this is short acting and averting an adrenal crisis requires ongoing treatment in hospital. A protocol supported by the AADAI and informed by the international panel of endocrinologists states that, upon suspicion of adrenal crisis, patients presenting with signs of extreme adrenal insufficiency should be treated with 100mg hydrocortisone IV or IM then 50mg every 6 hours plus a saline drip and undertake certain blood tests.

Part of the complication for medical professionals is that patients with adrenal insufficiency will present with different symptoms and at different stages of an adrenal crisis. Some hospitals and



some state ambulance services have protocols for treating patients presenting with adrenal insufficiency who are at risk of an adrenal crisis.

Evident across several stories were instances where hospitals failed to have adequate protocols in place for providing the necessary emergency response (Stories 4, 5, 6, 7, 8), or paramedics or the 000 call centre failed to appreciate the urgency (Stories 4, 9). Neglect occurs when protocols are not followed, symptoms and concerns of the patients are ignored, and the right people (endocrinologists) are not consulted to provide the correct treatment plans.

Some experiences of neglect during emergency treatment include:

- Paramedics are unaware of the protocols (e.g., the M24 Protocol in NSW) or they are not followed correctly, leaving the patient vulnerable to adrenal crisis (Story 4);
- 000 call centre operators are not medically trained and do not appreciate the need for urgent treatment (Story 9);
- Paramedics and doctors focus on the comorbidities (such as diabetes) (Story 8) or other more well-known conditions such as COVID-19 (Story 7) instead of dealing with the adrenal insufficiency, especially when no adrenal insufficiency protocols for clinical practice exist for paramedics (the COVID-19 panic exacerbated pressure on the health system and became a distraction, especially during emergency care, increasing fear and lack of trust in the health system);
- No priority triage on arrival at the emergency department (Story 6);
- Hospital staff disregard the medical information provided by the patient, including the medical ID bracelet (Story 4), fail to contact the hospital's endocrinologists (Story 16) or the patients' medical support team (Story 5);
- Withholding of IV hydrocortisone injection (Story 6) and removal of personal Solu-Cortef® injection while being treated in emergency (Story 6); and
- Delayed emergency treatment (administering of the IV hydrocortisone) that can have delayed consequences due to the severity of the symptoms (Story 7, 8).

Examples from Member Stories:

Member Story 4

NSW Ambulance attended and were completely unaware of Addison's disease or the protocol they are supposed to have in place. They did not understand why the injection was necessary... [At the hospital] My medic alert bracelet was not examined at any time... I was eventually examined by an orthopaedic registrar, and had to explain what Addison's was, and why I had had the cortisol injection. I gave full details of my medications, advising what my base rate of medication was for my Addison's. At no time was any attempt made to contact a member of the endocrine team.

Member Story 6

(2015) I presented at the Emergency Department in the beginning stages of an adrenal crisis and was not administered my emergency Solu-Cortef injection. Even worse, my emergency injection kit was taken from me by the emergency staff in the ED so I could not even administer it to myself. This is the equivalent of taking away someone's Epipen or insulin. After the 2015 incident was investigated, I was assured that an alert would be put on my file



so that emergency department staff will be aware of my Addison's Disease. Unfortunately, I had similar problems the next time I attended.

(January 30) I informed the Triage nurse that I have Addison's Disease and am immune-compromised. Due to an admin error on the part of the hospital, I was not seen for 7 hours despite repeatedly presenting at the reception desk and informing the Triage nurses that my symptoms were worsening.

(29 May 2022) I waited over 4 hours to be seen by a doctor only to be told that I did not 'look sick enough to be having an Addisonian crisis.'

Member Story 7

I heard the paramedics give handover to triage and they were clear about my presenting need to be given IV fluid and IV steroids asap. But when I was put into the emergency bay I was left there for hours, when the nurses checked-in on me I kept trying to say I need IV fluid and steroids for my Addison's [and] that the food poisoning symptoms are bad but that's not the life-threatening bit.

Member Story 9

NSW Ambulance has a protocol for treating adrenal crisis, called the M24 Protocol. However, the call centre operator who, I was later told, is not medically trained did not listen when I tried to explain the reason why I needed urgent assistance. The paramedics finally arrived 2 hours after my initial call. By this time, I was in a very bad condition with increasing adrenal crisis symptoms. The paramedics had a good understanding of what was needed to stabilise my condition.

3.3. Neglect during hospital treatment

People with AD/AI frequent hospitals, some more than others. People with AD/AI can be admitted to hospital for prolonged stays to recover from an adrenal crisis or to deal with other seasonal conditions such as the flu or COVID-19, complications from comorbidities, surgery and day procedures, and other reasons. People with AD/AI are advised to stress dose prior to procedures, such as double or triple dose their typical oral glucocorticoid replacement for surgery, gastroscopy or any other procedure. Additional fluids and IV steroids might also be provided during the procedures or emergency stays (as indicated in 5.2).

In these circumstances, careful management of the adrenal insufficiency is needed to overcome or prevent going into adrenal crisis while in hospital. Management requires medical staff to have adequate knowledge of and appreciation for the seriousness of the associated stress and emergency treatment required. Neglect in this context involves complacency causing failure to communicate with the correct medical professionals and failure to follow through with protocols.

Common across the stories were instances where:

- A failure to communicate with the endocrinology team in the hospital or the patient's GP or endocrinologist, resulting in the patient needing to administer their own medication (Story 4)
- Unnecessary late scheduling of surgery requiring fasting and an inability to have water to take scheduled medication resulting in elevated risk of an adrenal crisis (Story 6); and
- Locking away medications in a cupboard when admitted to hospital and the problems associated with this (Story 14, 16).



Examples from Member Stories:

Member Story 4

I suffer from a number of rare health issues, but my Addison's disease caused the most problems for both myself and the staff at a hospital in Sydney's northern suburbs, who simply did not seem to know what it was.

My medic alert bracelet was not examined at any time, nor was there any attempt to telephone to check my medical information.

I was eventually examined by an orthopaedic registrar, and had to explain what Addison's was, and why I had had the cortisol injection. I gave full details of my medications, advising what my base rate of medication was for my Addison's. At no time was any attempt made to contact a member of the endocrine team.

I was self-medicating from my own tablets which had been brought from home, as no-one at the hospital seemed to know what to do with me. I was nil by mouth from approx. 8.30am, in pain and stressed, so needed to have additional medication to avoid an Addisonian crisis. (See 5.2) Had I followed his instructions, I could have gone into a coma and died. At no point was any mention made of the 'Reach' program which I now know the hospital has for patients who feel unsafe, as I clearly did.

Member Story 6

I was scheduled for a day procedure at a large hospital. Prior to my scheduled surgery, I had provided medical documents to the hospital about Addison's Disease. On the day of the surgery, I spent over half an hour with the admissions nurse going through my medical history and talking in detail about my Addison's disease. The hospital and medical staff taking care of me had all been informed and were aware of my medical conditions. As the procedure is performed under general anaesthetic, I had to fast and was not allowed any water even to take my medications. As a result, my last dose of medication was at 6am and I was not called in to surgery until almost 3.30pm. That is over 9 hours which is far too long between medication doses. By 3 o'clock in the afternoon I was feeling lightheaded [and] having trouble staying awake, starting to experience mental confusion and my muscles were shaking. These are familiar symptoms that I have had before an Addisonian crisis. I had to self-inject my emergency injection because the nurses could not do it and the doctors were busy in surgery.

Member Story 14

During admission, my meds were taken off me and locked away, as were my replacement emergency injections (that I did not receive back). I did not get my doses on time and I was subjected to many invasive tests that were not required...

I was admitted into a 'locked ward' because I required 'a high degree of nursing care', they commenced me on antipsychotics which made me erratic and very fidgety - the opposite of what I am usually like with Addison's. This was a scary time where I had no access to my meds and was reliant on the nurses giving them to me on time. I tried to explain how important this was but on numerous occasions they sent nurses to inform me that I had received the wrong dose.



3.4. Neglect during monitoring and ongoing treatment

Ongoing monitoring of AD/AI is complicated by the differences within the medical profession as to what drugs should be used, what monitoring tests to take, and how much time a doctor gives to their patients. For example, glucocorticoids hydrocortisone and cortisone acetate are preferred by medical doctors over prednisone/prednisolone; there are also differences in opinion as to how much (low physical dose or a more flexible dose dependent on patient needs) and what regime of drugs are taken across the day (once, twice, three or four times, or whether to move to a pump or to mix the drugs). Fludrocortisone (mineralocorticoid) is prescribed for people with Addison's disease. Some endocrinologists have more experience in treating or researching AD/AI so they bring those experiences to the patient-doctor negotiation of treatment. Solu-Cortef®, our life saving emergency injection is not always prescribed. There are few blood tests that indicate the 'correct' steroid dose for an individual, though a number of tests are used, along with other tests to look at other effects. Endocrinologists are sourced by people with AD/AI from public or private hospitals or medical clinics, with some endocrinologists more than others making time to explain, discuss, and work with their patients during consultations and be contactable out of hours for emergency situations.

A number of experiences have been portrayed through member support groups that describe feelings of frustration or distrust in their endocrinologist or doctor. Patient care is essential, especially while a person is finding their new normal after diagnosis. Negligence in this context involves a failure to recognise the need for close monitoring that takes into account how the patient 'feels' alongside the blood tests, failure to prescribe the drugs needed, failure to recognise the education needed by a person new to AD/AI, and/or a failure to acknowledge the lived experience of patients.

Across the stories there was evidence of the following:

- Some doctors take a 'textbook' approach to prescribing the regular dose of steroids, without appreciating the need to adjust the dose to suit the individual (Story 14). This can occur in hospitals or when the patient does not have a regular endocrinologist or doctor who they trust.
- Some medical professionals do not prescribe Solu-Cortef® because they don't believe patients know how to administer it properly, or they believe in an emergency an ambulance can arrive in 20 minutes in the city and they can administer it. However, not prescribing Solu-Cortef® to an adrenal insufficient patient is likely to be regarded as negligence by the medical board (person comm. endocrinologist, name withheld), especially given that ambulances in some states do not carry hydrocortisone and the current ambulances delays across Australia. Training to administer Solu-Cortef® should be provided by a medical professional; but if they do not, training videos are available on the internet and through the AADA website.
- There were some instances where prescription drugs contraindicated the drugs for AD/AI, such as a migraine medication contraindicating hydrocortisone. This requires patients to be self-educated.
- A general dislike by the medical profession of patients telling them how to treat adrenal insufficiency or adrenal crisis (Story 1), or self-help or support groups that might provide

incorrect or different information. As a self-managed disease, people with AD/AI need to be self-reliant and must be self-educated.

Examples from Member Stories:

Member Story 1

I keep reading from my notes, 'injecting hydrocortisone "is safe", even [if] out-of-date or not needed, and 'it can't have serious or long-term side-effects, according to Professor Wass, in the United Kingdom...'. Dr Baby's interrupts with his usual squeal, 'Was that on the internet? Anyone can find some crazy person on YouTube to say whatever you want them to.'

...

'And shouldn't I be on hydrocortisone 200mg/day continuous IV for 24 hours?' Dr Baby starts up again, 'That's only for adrenal crisis, and you aren't in any type of crisis.' 'I was,' I reply – immediately annoyed at myself for taking his bait. 'No, you weren't,' retorts Dr Baby. I politely respond, 'Your hospital's "Plans" say the medical staff are to chart hydrocortisone, even if, "the patient suspects a crisis".' Dr Baby replies, 'I'm the doctor. Not you. So, I am the one who is trained for, and responsible for, deciding what medication I prescribe, no matter what patients may ask for or what they may think they want.'

Member Story 14

I believe I needed gradual tapering and consistent levels of hydrocortisone (probably at a stress dose due to emotional stress) ... They discharged me with a 7-week weaning schedule down to 20mg a day. I was not feeling at all well at this dose.

On return to the UK ... I explained that the doctors had put me on a weaning schedule but that I was not doing well at all on it ... Endocrinologist A said 'we have increased her hydrocortisone doses today to 15mg/15mg/10mg as her 4 hour and 8-hour cortisol values are below our target of 200-250nmol/L giving us biochemical evidence that her current doses may not be adequate.'

3.5. Neglect during the Diagnosis Odyssey

"A diagnostic odyssey is defined as the time between when a symptom or feature of a genetic or rare disease is noted to the time when a final diagnosis is made."

FDNA Teleheath (FDNA.com)

Symptoms of adrenal insufficiency may include:

- Loss of appetite, nausea, vomiting, diarrhoea, weight loss, abdominal pain
- Fatigue, weakness, headache, dizziness (especially on standing)
- Difficulty concentrating, mood swings, depression
- Salt cravings and darkening of skin and mucous membranes (in Addison's Disease)
- Irregular menstruation in women
- Low blood pressure



The 'odyssey' is that sometimes people can be 'running downhill' – feeling nauseous, notice they develop a bronze skin tone, crave salty food, lose weight and muscle tone – but do not get tested or receive a diagnosis for Addison's disease (primary adrenal insufficiency).

With Addison's disease in particular, symptoms are often missed or misdiagnosed due to the symptoms being similar to many other diseases. Treatment is ineffective because the root cause of the symptoms is neglected. Nausea and vomiting are given relieving medications without making the connection with other coexisting symptoms. Fatigue arising from diminishing cortisol is mistakenly treated as chronic fatigue. Anxiety and depression are treated as stand-alone conditions when they are actually caused by Addison's disease.

The cost of these tortuous diagnostic odysseys can be very significant both financially (living in 'genteel medical poverty') and socially through relationships that never eventuate or collapse under the strain. The draining effort to find the correct diagnosis as people go to different specialists can impose very real mental health costs.

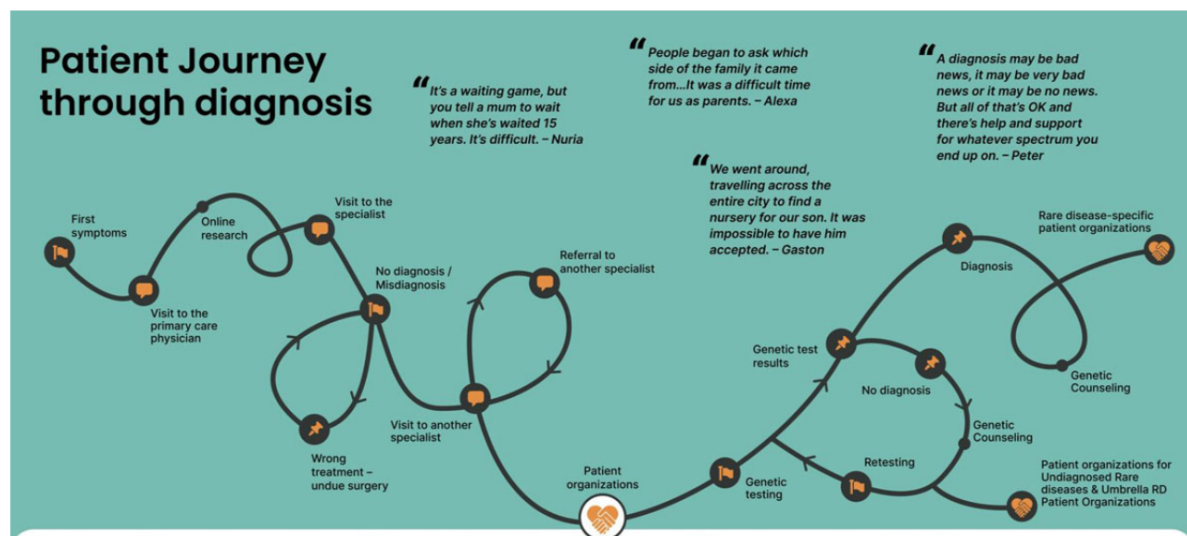
The patients included in our submission lived to tell their stories. To have lived so long, they may have had some residual production of natural cortisol before encountering a deep crisis. Subclinical adrenal insufficiency is documented in the medical literature as a stage before clinical intervention is required. It helps explain the duration of some odysseys, but the AADAI's concern is that recognition of a rare disease as a diagnostic possibility is essential so that the period spent in the clinical stage is not extended needlessly.

There are enough horror diagnosis stories in our submission to call for systematic improvements in awareness of rare diseases being at least considered among the causes of the presenting symptoms. We need to learn from the errors demonstrated by the extreme cases that went to the Coroner's Courts (Albany WA and Sydney NSW) to ensure they are not repeated.

Statistics relevant to the diagnosis odyssey experience:

- Incidence of primary adrenal insufficiency (Addison's disease) – 8 NEW cases per million population/year in first world countries
- Incidence of secondary adrenal insufficiency – about 15 NEW cases per million population/year (variable)
- Prevalence of primary adrenal insufficiency (Addison's disease) – 1 living case per 8000 population in first world countries
- Prevalence of secondary adrenal insufficiency – 1 living case per 5000 population in first world countries
- GPs average a little over 1 to every 1000 population in Australia. Thus, a GP is unlikely to be dealing with one of the 200 new cases in any single year, but there is a 1 in 4 chance they will be treating a patient living with Addison's disease and every second GP is likely to have a patient with a type of adrenal insufficiency.
- It takes on average four years to reach a diagnosis (Medics 4 Rare Disease Training program)





(eurordis.org/publications/solve-rd-infographic-on-the-patient-journey-to-diagnosis/)

Some observations by members of inadequate diagnostic attention by medical staff include:

- Delay in achieving a correct diagnosis resulting in more severe illness and longer recovery time (Stories 2, 7, 10, 11, 12, 14, 15)
- Delay can be costly – in actual cost of treatment, in loss of income if forced to resign, in estrangement from friends and family (Stories 2, 13, 14, 16)
- Being in the care of a medical practitioner who recognises the need to refer unusual cases on can be a matter of luck. The chances of having such a doctor in rural or remote areas may be low but also applies in large population centres and even in hospitals. (Stories 10, 12, 13)
- Assumption that a mental health condition in the patient explains the failure to find a physical cause of symptoms (Stories 2, 7, 13, 14)
- Alternative physical causes become fixed in the diagnostician's mind at the exclusion of the real cause – adrenal insufficiency. It is an extremely dangerous assumption because the resulting adrenal crisis is a life-threatening condition (Story 7 [Incident 1] and Stories 13, 14).
- A combination of the previous two points where the patient is accused of not 'owning' a pre-existing condition (diabetes) rather than believing the patient and acknowledging the reality of adrenal insufficiency symptoms (Story 7)
- The failure to get well can cause anxiety symptoms (Stories 7, 10)
- A vital clue is missed in cases where the patient is given a corticosteroid to treat the 'other' condition, and suddenly they are much better – only to fall ill again when they are weaned from the corticosteroid. Instead of this observation prompting the medical staff to investigate the patient's cortisol levels, there is an unexplainable reluctance to consider adrenal insufficiency – even among some endocrinologists (Stories 2, 13, 14)
- Reluctance to perform an ACTH stimulation (synacthen) test when it would show presence and type of adrenal insufficiency (Stories 13, 14)
- Not feeling 'listened to' by their treating doctors and observing that advice from fellow medical staff including those with specific expertise seems not accepted (Stories 2, 7, 13)

Examples from member stories:

Member Story 10

The saying, 'never give up', seems cliché. However, there are a few things you need in order to not give up. You need an Endocrinologist that believes in you; that's in your corner. You need a family or an advocate that believes in you, someone that will protect you when you are so sick that you can't even make sense. And a great support group.

Member Story 7

The doctor assumed that I hadn't made peace with my diabetes diagnosis, she said to me: 'You just have to make a choice and get on with your life...you can't let diabetes own you...I have a friend that has Type 1 diabetes like you and she just ran a marathon...she doesn't let it get her down, you need to be tougher.'

...

I knew something was wrong, but all I could manage to say to the doctor was that 'I didn't take any pleasure in being in "a sick role" and that I don't like hospitals', I told her 'I was a proud woman and if I had to get help to get to the toilet and wipe my bum then something was wrong.'

Member Story 12

I was in the Infectious Diseases ward with salmonella, and (the official description) 'a wasting disease'... By the next morning I was in hospital, the available medical staff were unable to diagnose my underlying problem. I had concluded that I was probably going to die. I had a visit from a Tamil doctor who was interested in the medical facility. He examined my notes, asked me a few questions (including, with a trace of a smile, 'Do you have Indian blood?'), and left the room for a brief discussion with the local endocrinologist. Not long afterwards, the endocrinologist re-appeared to announce that the mysterious 'wasting disease' had been identified, and could be treated.

Member Story 13

I kept on coming back to stories about people with Addison's who felt better once they were hydrated and had salt and I said to my mum, that's me. I'm 40 years old and I weigh 42kg. My GP did a strange thing, he referred me to a very good physician. By this time, I had an endocrinologist and was diagnosed with Type 1 diabetes and then Type 2 because of the steroids. Then I had Cushing's because I had too many steroids, as I needed the prednisone to control the anaphylactic shock. I also had an immunologist now, called Dr K. He too had no idea why this was happening and why I felt so well on prednisone. IVIG was prescribed for my immune system and the myopathy. It was wonderful. For the first time I had energy, I could stand up, I wasn't throwing up. And then they decided to wean me down off the prednisone and that's when the problems started again.

3.6. Reporting neglectful treatment

It is important that patients feel they can influence the quality of care they receive. When incidents are reported to medical professional bodies, ambulance services and hospitals, it is an indication that patients have experienced what they consider to be inadequate care or neglect, some of which have been outlined in the above themes. While many people with AD/AI experience some of these issues, few choose to or are in the right frame of mind to report negative experiences.



Best practice should involve both an easily accessible reporting system for patients as well as timely completed incident reports by medical professionals when an AD/AI patient fails to receive adequate care. Both are needed to ensure professional and patient perspectives are reviewed for effective change.

Evident in some member stories are instances where, in response to patient feedback or complaints, medical organisations do not provide adequate answers as to why the AD/AI patient has endured poor treatment. Members have expressed some frustration with struggling to navigate the formalities of cumbersome reporting systems. Medical organisations fail to share if or when they will implement an effective process or protocol for future patients with the same medical condition, and then how this information is to be shared with the public. The stories highlight the dismissive nature of medical professional organisations that fail to provide an answer for why appropriate treatment and care are not always provided in government medical settings.

Stories highlight:

- A cumbersome reporting system (such as many emails and phone calls) that caused a delay in reporting an incident and further delay in receiving answers from the NSW ambulance service. The response received was dismissive and suggested that, in the event of an emergency, the AD/AI person should lie about the type of condition (heart problems) they have in order to be triaged to receive immediate ambulance care. (Story 9)
- The psychological barriers to reporting neglect, including a fear of 'not being taken seriously', and that patients need support to be able to share their stories and locate the necessary processes to do so, and that a risk analysis report should be made by the attending medical practitioner in a critical near-miss incident. (Story 7)

Also evident are instances where hospitals have responded positively to patient feedback, sometimes in response to the first letter or complaint, or in response to a follow up complaint. Sometimes a mediation meeting resolved the issue. Positive outcomes experienced included written and/or verbal apologies, letters of explanation, and/or description of changes to be implemented. Some examples include:

- A reply letter explaining that new protocols and systems would be shared with hospital staff and visiting professionals. However, while the protocols were new, they were not shared with the member or shared publicly. (Story 5)
- A patronising reply letter, but when followed up again, an invitation to attend mediation, leading to an apology and a clear outline of the many changes that were to be implemented in the future. (Story 4)

Examples from Member Stories:

Member Story 4

After receiving a very patronising letter from the Director of Medicine, Dr K, in May 2019, which essentially implied I had misunderstood, been confused by [the] anaesthetic, or was just plain wrong, I wrote back reiterating the issues, and was finally persuaded by HCCC to attend mediation with the hospital.



On entering the room, I received an immediate apology from Dr K who said the letter was totally inappropriate, offensive and did not address the concerns raised. He confessed he had been on leave for 3 months and been pressured into signing the letter on his return without really reading it as had taken so long to reply. This set the tone for what proved to be a fruitful meeting.

This apology was repeated in a letter subsequently received, outlining the steps the hospital is now taking to improve the situation for any Addisonian attending the hospital emergency department.

Member Story 7

I believe that the doctor's bias towards me put my life in jeopardy, she effectively discharged a patient who was on the verge or possibly already by then in organ failure... Why haven't I reported it officially till now? I have been wanting to report this since it happened, but every time I sit down to write it, I feel scared that I won't be taken seriously. I told my family and friends about the incident, and some months after the Addison's diagnosis I started seeing a Health Psychologist due to my psychological distress regarding my mistreatment and overall difficulty with my physical and psychological adjustments to now living with two life-threatening chronic conditions. Four years later and with much support from friends, family and my psychologist I am finally in a place where I can find my voice to share this experience in written form, even though I am still working on healing from the trauma.

...

I want to know if a clinical risk incident was submitted for a near miss pertaining to my medical event. Leaving me for hours in triage without an endocrinology consult and consequent medical treatment in terms of administering steroids, [that] put me at a potential life-threatening risk.

I want to know if a clinical risk incident was submitted then what reviews and corrections have been made to policy and procedure to ensure this type of missed step doesn't occur again? Was this a case of medical neglect in that a staff member breached protocol by not contacting endocrinology for consult? Or was this a matter of not having a clear policy on adrenal crisis management, a problem with procedure, or other?

I recommend that if hospitals are going to put posters on the walls stating that they are patient-centred and our voice matters, that all staff are clear on how to support patients to give feedback. What the processes are, where the forms are kept and how to submit them or who to talk to.

Member Story 9

In February, 2021 I had an unfortunate experience with NSW Ambulance Service.

...

I live alone so I rang for an ambulance at 4pm as I had food poisoning, meaning my medication could not be absorbed. NSW Ambulance has a protocol for treating adrenal crisis, called the M24 Protocol. However, the call centre operator who, I was later told, is not medically trained, did not listen when I tried to explain the reason why I needed urgent assistance. The paramedics finally arrived 2 hours after my initial call. By this time, I was in a



very bad condition with increasing adrenal crisis symptoms. The paramedics had a good understanding of what was needed to stabilise my condition....

I tried to follow up this letter on a few occasions but was unable to contact the Executive Director, Clinical Operations, NSW Ambulance, as advised in the letter. The two concerns I wanted to raise were the long wait time of 2 hours in a life-threatening emergency and how could I be flagged in the system so this does not happen again if I need an ambulance?

It took me 3 months, with many emails and phone calls, to finally reach a resolution to my situation. I was actually told that, if I needed to call an ambulance again, to say I have chest pains or breathing difficulties and this will get me a faster response. As a truthful person who does not like to 'queue jump' I would be most reluctant to take this advice.

The eventual outcome was that I am now 'red flagged' in the NSW Ambulance system. I had to give permission, due to privacy concerns, for my medical condition to be displayed in their system, which I was happy to do.

Unfortunately, I feel I can no longer rely on, or have trust in, the NSW Ambulance Service. I have had to make alternative arrangements with my family and GP to get emergency care should I need it again. This has also impacted my mental health, with frequent flashbacks, and this causes me anxiety.

4. Recommendations

Many of the member stories included recommendations, including 'what should have been done'. We have distilled these into four core recommendations.

4.1. National approach to emergency management

A National approach to emergency management for people with acute adrenal insufficiency and adrenal crisis based on best practice must be introduced to 000 call centre operators, ambulance services, other emergency services, and emergency departments.

A national approach would reduce replication of effort in areas such as the following:

- A. A common approach to emergency management when presenting at the hospital emergency department, for example:
 - a. Individuals being flagged in the local hospital system to support efficient and appropriate triage
 - b. Hospital protocol in the system (triggered by the registry) that is initiated at triage and then the steps of the protocol followed. The triage system needs to prioritise people presenting with suspected adrenal crisis at the highest level.
 - c. Pay attention to individual's medical plans as each person presents differently; paramedics and doctors in hospitals need to look to more than medicine but also the ongoing steps needed.
 - d. A national rare disease registry where AD/AI patients' details can be accessed by emergency staff.
- B. A common approach to call centre operations (000) that includes red flags for AD/AI.
- C. In relation to ambulance and emergency:
 - a. A national approach to ambulance officer training in emergency medicine.
 - b. Consistency in the national approach to the provision of ambulance services for AI/AD patients throughout Australia.

4.2. Education for medical professionals and allied health professionals

Education should be provided to medical professionals and allied health in relation to treatment options and the risks of acute adrenal insufficiency and adrenal crisis. This includes doctors, surgeons, dentists, nurses, paramedics, endocrinologists, mental health practitioners (psychologists, counsellors), and endocrine nurses. Education can be provided at all stages of a medical professional's career. Without a shift in how medical training is delivered, this will be challenging, but when considering that AD/AI requires careful emergency management (compared to some other rare diseases) and the dire consequences if symptoms are not recognised, acknowledged and treated adequately, efforts need to be made to ensure medical professionals are adequately educated.

Education can focus on the following:

- Raising awareness of AD/AI crisis through information from peak bodies and patient groups and associations.
- Understanding the dangers of unconscious bias in medicine in addition to any professional communication training that focuses on client centred interviewing for assessment and empathy (Story 7).



- Education and clear process to avoid perceived barriers for staff to escalate concerns to senior medical staff. Hornsby Hospital in NSW, for example, is educating its staff about the 'Speaking up for Safety Scheme,' which encourages staff to seek help when faced by such issues (Story 4).
- Team approach to diagnosis, ongoing medical treatment, and a shared responsibility for education: "You need an Endocrinologist that believes in you; that's in your corner. You need a family or an advocate that believes in you, someone that will protect you when you are so sick that you can't even make sense. And a great support group." (Story 13)
- Development of better tools to support diagnosis and treatment for doctors in hospitals and GPs.

Some examples of medical professional education already available:

- The Rare Care Centre in WA is introducing post-graduate rare disease training for nurses through Curtin Uni. (<https://pch.health.wa.gov.au/Our-services/Rare-Care-Centrefabulous-centre>)
- The Medics 4 Rare Disease Australian modules are currently being developed and have specific information for medical professionals. Backed up six weeks later by ECHO Zoom sessions where the module content is discussed.
- The 'Rare portal' by Rare Voices Australia (<https://rarevoices.org.au/rare-awareness-rare-education-r-a-r-e-portal/>). See also the National Action Plan for Rare Diseases (<https://www.health.gov.au/resources/publications/national-strategic-action-plan-for-rare-diseases>)
- The introduction of and training in using Clinical Practice Guidelines for dealing with adrenal insufficiency by the Victorian Ambulance Service during 2022, which have made a huge difference to the way Victorians with AD/AI receive care.

4.3. Showcase best practice

Showcase best practice that incorporates GP/specialist diagnosis, GP monitoring and wholistic care/team approach to treatment that includes endocrinologists and endocrine nurses. We are calling for best practice to be established by the medical profession, in consultation with and informed by the lived experience of people living with AD/AI, and which captures the whole cycle of diagnosis, monitoring and management, and emergency management, and the diverse presentations that doctors might encounter. A point of neglect has been that doctors can defer treatment until adrenal crisis is clearly present, rather than act to prevent adrenal crisis (Stories 1, 4). Member Story 17 provides contrasting stories, one which might be considered best practice, another which may reflect the pressures that medical staff are under that can result in neglectful treatment.

Best practice can focus on, for example:

- Triage AD/AI patients in an adrenal crisis as emergencies from when an emergency call is logged;
- General Practitioners and emergency staff to recognise that AD/AI symptoms may present as other illnesses and consult with the patient's advice or as outlined in their medical care plan before assuming and prescribing treatment and medication;



- Encourage a whole team approach to communication between ambulance staff, emergency staff and specialists to ensure appropriate treatment is given to the patient while in hospital and when they are discharged; and
- Processes for dealing with complaints, for example, design a simple medical neglect incident reporting system for patients.

4.4. Collaboration between professional and member associations

Professional associations (e.g., Endocrine Society of Australia, Endocrine Nurses Society of Australia, ANZSPED – formerly APEG) work with peak bodies and associations (member associations) such as AADAI and RVA who advocate for and support their members and communities with rare diseases. This should be a dialogic relationship where these member associations communicate lived experiences to professional associations, and professional associations support the work of the member associations by co-designing protocols and best practice, providing and listening to advice, answering questions, and undertaking research.

Member Story 1

The Worst adrenal crises were Not the Scary ones

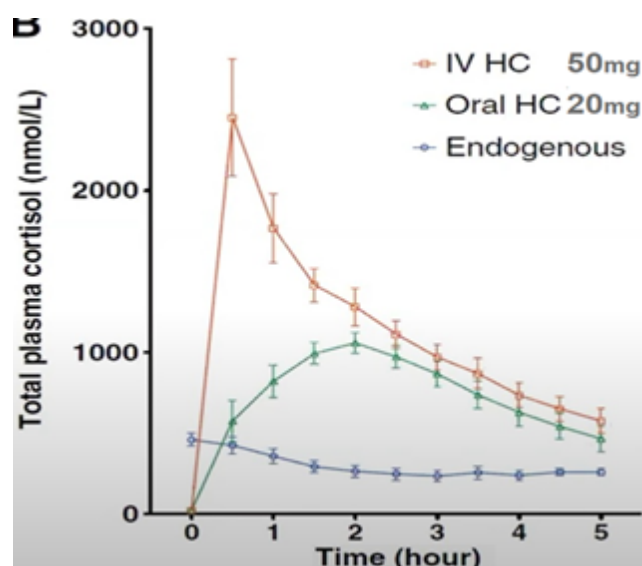
May I assume we know to layer protection around ourselves? Alert bracelet; up-dose early; train yourself (and others) to inject; written plans for yourself, the ambulance (not least to avoid triage nurses and waiting rooms), Emergency Department (ED), hospital ward; etcetera. Did I forget any? I suppose I assumed – wrongly – naively – following the rules magically makes everything go well. After 3 years and 25 adrenal crises (AC), the most stressful and scary weren't the severe ones – far from it. That still seems more confusing than helpful.

My severe crises were different: sudden and obvious: hours of heart rate double (120bpm) my usual (60bpm); or bad diarrhoea; or bad vomiting, even if I took a sip of water; or colourless skin; or delirious; or low sodium and glucose; or obvious injury or illness (e.g., gastro germs). Each time, doctors showed no doubt. They treated me quickly – and kept treating me until I bounced back (or improved over several days). Even when doctors describe crises as severe, frankly, they didn't induce worry or trigger significant stress

Dad's friend Paul was a B-17 bomber pilot in WWII, who flew bombing missions over Germany with other aircraft, including the 'Memphis Belle'. In the early 1990s the three of us watched a movie of the same name, based on a true story. Paul said, the emergencies were accurate, but it was ridiculous to squeeze 3 years' worth of problems into a single flight. So, as I acknowledge that I'm being ridiculous, would you permit me to stir in 3 years' worth of issues (and mixed metaphors) to brew a fictional perfect storm of adrenal crisis unpleasantness, based on a true story? If so, I'll try to show how I got myself stressed, annoyed, and into lots of trouble, despite doing a few things 'correctly'.

Here goes: I'll try to dream up my fictional, Nightmare on Horror Street. Infection (e.g., gastro) and stress commonly trigger crises. I'll pick stress, since that should make doctors are less likely to believe me. Stressing, sweating, and peeing a lot isn't great. I'll up-dose fludrocortisone and eat spoons of table salt. This will push my sodium levels just into the 'normal' region and helps the doctor (prematurely) to exclude AC. I don't want major or clear symptoms, since that will help a doctor to believe me. Fortunately, AC signs may be non-specific. You may even look well. So, I'll choose horribly vague "feelings": cold; dizzy; drowsy; fatigued; weak; light-headed; nauseous; urinating too much; irritable; and confused. (Yet, if I am confused, I'm likely to deny it.) I'll follow my endocrinologist's advice: inject hydrocortisone and call the ambulance. Ms Paramedic arrives 20 minutes later. One must always be honest. So, I say my vague symptoms are gone now. Ms Paramedic follows her rules, giving me a litre of IV saline. We drive to ED. And wait. An hour after injecting, I feel okay. My cortisol levels are probably great (see graph.7 Thanks to the recent AADA seminar).





Ms Paramedic reports (accurately) to Ms Tired-Nurse, 'Ben feels okay. He complained, but we didn't see any symptoms.' Ms Tired-Nurse laughs while reporting to Dr Busy, 'Bed 1 reports no symptoms, and that's what Ms Paramedic saw.' Dr Busy says something rude, then ignores me. Dr Busy tells 22-year-old Dr Baby to take a break from Facebook. Dr Baby ignores all I say, and tests me for covid, twice. It's negative, twice. Finally, 3 hours after I injected myself, I try 'politely proactive'. 'My endocrinologist told me, "You don't need to look sick to go to ED If you look sick, you're probably too late and already in big trouble"'. Ms Tired-Nurse decides I'm lying, stupid, crazy, or all three. Doctor Baby, now satisfied I don't have covid, says 'I'm discharging you.' 2 minutes later he says, 'if you conjured up some feeling that you know more about medicine than the amazing professors who have taught me full-time for the past five years, you're more than welcome to phone and talk with the off-duty nurse about it. Bye.' I'm still polite, compliant, confluent even as a security guard pushes the wheelchair out to the waiting room. That is, confusion overtook my self-awareness, and I'm unable to phone anyone, or to argue, or even to notice the warning signs. Some hours later, at home, my wife wakes me up. I feel death approaching. She calls the ambulance. Mr Medic fixes me up. The next thing I know, Mr Medic hands over to Nurse Exhausted, 'saline... hydrocortisone...', something else. Nurse Exhausted talks to me. She listens out of habit, or because she feels obliged. 'The experts say, 'prevention is key' with adrenal crises, I offer, politely. 'I wish that was how A&E worked, sweetheart,' she replies, honestly. Oh, no. Dr Baby returns. His face is the angriest shade of crimson. He awkwardly pats my shoe while failing to pull off a jovial tone, 'I thought we discharged you?' That failed too. He switches to a serious tone. 'Anyway, I'm going home. And deep down, I know that you know, that you should go home too. Right?' My bed faces a clock. It says, 9pm. I can't remember if I arrived here at 6pm, or that's when Mr Medic injected me. Dr Arrogant comes over. He cared less than nothing for what I said. I phone Dr Endocrinologist and hit the speakerphone button. He tells Dr Arrogant to inject hydrocortisone. 'Okay, Dr Endocrinologist. I'll do that right now,' says Dr Arrogant.

Dr Awesome walks over. She has the best smile ever. 'Hi! I googled Adrenal Crisis and learnt a lot. Can we chat?' 'Yes,' I reply. 'If we don't treat you, you might die. But, treating you is easy, almost zero risk, and you'll probably be back to normal in a few days. Sorry to be so dramatic, but Australian law requires me to tell you about the risks of different treatment options, risks of not treating, risks specific to you with adrenal insufficiency – in fact risks you probably care about, like dying. Does that

make sense?’ ‘Yes,’ I reply. So, Dr Awesome goes on. ‘You’re really dehydrated, and you’re at risk of an adrenal crisis. So, I think it is best, for the first day, if we give you lots of cortisol, around 200mg, and we check on you regularly, especially to ensure you get enough fluids. The risk is extremely low – almost nothing. I don’t know of any better option. Or, if we don’t do this, or if we delay, there are huge risks, such as: blood failing to circulate around your body properly; heart failing; other organs failing; coma; and death. I suggest treating is better than not treating. Does that make sense?’ ‘Yes, perfectly clear,’ I reply. Dr Awesome continues, ‘Clearly, you understand the risks and you’re capable of deciding. I’m not allowed to force you to choose the option I want. That is, the choice is yours, not mine. ‘Let’s start the treatment option then, please, Dr Awesome?’ ‘Oh, no,’ she replies. ‘I’m not real.’

She disappears as I realise a man is shaking me. My racing heart seems to be pumping something like pavlova. The ED clock no longer says 9pm; now it says midnight. The room is jumping around. No. It’s my body that’s bouncing around. Dr Arrogant is shaking me, violently, but I can hardly feel it. Why does he look so unhappy? ‘I can’t hear you!’ he yells at me again, and again. ‘Are you okay?’ he yells. I try to speak but I can’t hear myself. I’m making no sound. He’s bouncing my head up and down into the pillow. I want him to stop. I try to grab, but my arms aren’t moving. The bed is tilted up. I see dozens of electrical leads draped over the bed rail – not connected to me. An air hose leads to a blood pressure cuff that is inflating by my side – not on my arm. My other arm is cannulated. I recall the paramedic did that. There is no IV line in there. I’m not connected to saline. I’m not connected to any machine. Have they all just ignored me for 3 hours? Dr Arrogant is still whinging at me. Can’t I die to something better than this? Anything is better than dying with Dr Arrogant angrily shaking me that my death is interrupting his flirting with the nurses. Oh, good. He’s gone. No, he’s starting to freak out; to yell at a nurse; oh, and to yell at me – in case I’m now partially deaf? ‘You’re... having... an adrenal... crisis! I’m... injecting... enough... cortisol... to wake up... a dead horse!’ Now I’m wondering what that means. I pass out, again.

Dr Baby wakes me up, ostensibly to ask if I ate breakfast. I ignore the question in my mind – would I rather dying with Dr Arrogant or Dr Baby? I politely ask why he discharged me yesterday, instead of treating me ‘without delay’ as it says in ED plan written by his boss, the ED director. And, the experts say that cortisol is essential for survival; it’s vital for survival, homeostasis, etc. And, adrenal crises can progress very, very quickly. So, don’t wait! Half an hour is too long. And, if the question of a crisis even comes into your mind, or if doctors are in doubt, inject! ‘And, ‘clinical suspicion requires immediate’ Hydrocortisone. Admittedly, I should have stopped reading from my notes after the first sentence, but I wanted to reinforce my point. Dr Baby replied, ‘there was no doubt. There was no suspicion. You weren’t in crisis.’ I almost open my mouth but recall that adrenal crisis has no widely accepted definition, so there’s not point me arguing that point. 22-year-old Dr Baby digs deep, putting on his most condescending voice, and dropping an octave, ‘in professional medicine, we medical practitioners are required to consider literally thousands of factors, such as you manifesting no symptoms at all, and weigh those against thousands of other facts such as the extremely serious risks associated with administering unnecessary medications to perfectly healthy persons.’ I keep reading from my notes, ‘injecting hydrocortisone ‘is safe’, even out-of-date or not needed, and ‘it can’t have serious or long-term side-effects, according to Professor Wass, in the United Kingdom...’ Dr Baby’s interrupts with his usual squeal, ‘Was that on the internet? Anyone can find some crazy person on Youtube to say whatever you want them to.’ He pauses to engage his deep, fake voice again, ‘Notwithstanding, healthcare providers know that healthy people shouldn’t tarry in hospital as you have a much higher chance of contracting a serious infection... and this is even more true for

patients who do actually have underlying medical conditions. By now, you must agree that it's best for everyone, especially you, that you go home now.' Ignoring his emotional manipulation, I check I'm still calm, polite, and working systematically through my checklist. 'Aren't we all supposed to follow the ED Director's plan? e.g., to ensure I'm stable on oral steroids before discharge?' Again, I should have let him answer, but I want to make the most of him not yet interrupting me. 'And shouldn't I be on hydrocortisone 200mg/day continuous IV for 24 hours? Dr Baby starts up again, 'That's only for adrenal crisis, and you aren't in any type of crisis.' 'I was,' I reply – immediately annoyed at myself for taking his bait. 'No, you weren't,' retorts Dr Baby. I politely respond, 'Your hospital's Plans say the medical staff are to chart hydrocortisone, even if, 'the patient suspects a crisis.' Dr Baby replies, 'I'm the doctor. Not you. So, I am the one who is trained for, and responsible for, deciding what medication I prescribe, no matter what patients may ask for or what they may think they want.' Dr Baby is clearly sick of my patter, so he starts reading my notes over my shoulder. 'Delays can kill. Untreated AC is fatal. AC 'is an endocrinologic emergency with a high mortality rate, and requiring immediate recognition and treatment to avoid death. Its clinical course is unpredictable; progressing quickly to death; often need ICU; Hydrocortisone 200 mg/day continuous IV is best (boli doses fail to mimic circadian secretion). Inflammation, tissue damage, and bacterial infections are more common in adrenal insufficiency patients. NSW clinicians with less diagnostic ability / experience / in smaller rural hospitals may be unfamiliar with AI needs / fail to treat quickly or adequately.¹⁵ Ignorance of AC as emergency can result in Px communicating the need for urgent treatment to hospital staff but ignored or treatment refused with grave consequences. Dr Baby speaks again, 'Yes, all that's true,' his chest puffed out as if medical researchers always ask for his advice before they publish.

Somehow, hours later, Dr All-Talk wake me up. We're in Medical Ward. He promises to chart / prescribe saline and hydrocortisone, according to the hospital's official plan. The next morning, I wake up, heart feeling like it's pumping pavlova. I can't speak. I can't move. Really? The nurse explains Dr All-Talk charted nothing at all. Dr All-Talk returns that afternoon, assuring me it won't happen again. The next morning, I wake up, heart feeling like it's pumping pavlova. I can't speak. I can't move. Really? Again!

Member Story 2

Diagnosis:

The tale of my eventual diagnosis is a long and winding road. Looking back, I had the warning signs of adrenal problems throughout my childhood, but vomiting episodes were baffling to the doctors and it was eventually termed “migraine stomach”. Any stress/stressor brought on these episodes and I got to dread even the childhood delights of holidays and invitations to birthday parties etc. My parents assumed it was rich food at parties that triggered things. In my teens and competing in athletics events locally and nationally, the vomiting episodes were put down to competition nerves.

Things seemed to improve in my early adult years, with the occasional “atypical gastro” attacks. When doing further study in my late 40’s and 50’s, I went to a local naturopath for a pick me up as I was heading to the USA for several months to study and was fatigued. He was visibly shocked when he diagnosed adrenal insufficiency and prescribed some herbs. Unfortunately, I had no idea what adrenal insufficiency was and he did not elaborate. I can remember on the long flight to the US I felt so ill I began to think that I would not make it to the other end, but luckily after some food and a long nap I rallied.

Not long after, I developed a goitre and my then doctor prescribed thyroxine 100mcg and also sent me to a nurse he knew for colonic irrigation to help with bloating. Apparently, that was an unusual high dose first up and after attending the colonic irrigation the same afternoon I started the medication, I collapsed and was rushed to hospital in an ambulance. My body obviously couldn't cope with the loss of electrolytes as well as a large dose of Thyroxine. A friend who was with me at the time said that nothing the doctors did seemed to help until they gave me IV hydrocortisone and I rallied. None of this was relayed to me by the hospital, or my then doctor. I now realise I had my first adrenal crisis. There followed several episodes and I was virtually an invalid for several months. The doctor proscribed prednisolone as he said I would be unable to function normally without it. Still no mention of adrenal insufficiency, or Addison's Disease.

After one ED admission, the hospital booked me in to the outpatients endocrinology department and I had my first appointment with an endocrinologist. He wanted to do a Synathcin test and changed the Prednisolone to Cortate. It was some time before I felt up to having the Synathcin test and this wasn't helped by another doctor telling me that having the test was worse than the disease!

Eventually I had the test and was told I had Addison's disease and discharged from hospital and I felt as though I had been through several crises in one day. Unfortunately, whenever I ended up in the ED after that, I was told that my test was borderline and that I didn't have Addison's. I have a feeling that the endocrinology teams were miffed that a GP had put me on Prednisolone without referring me to a specialist. They told me that my adrenals were struggling because of the period I had been on the steroids. They just wouldn't listen that it was the fatigue and episodes that preceded the steroid use.

About two, or three years later, I had to report to AHPRA one of the GPs who had earlier treated me (later suspended for several months) and surprisingly, at this time my endocrinologist completely reversed his opinion and said that he now considered that I could well have Addisons Disease. He



then signed me off into the care of my then GP who specialised in hormonal problems. I considered at the time that perhaps he considered I might be a problem patient and best be rid of me. Luckily this new GP had a good grasp on adrenal and thyroid problems and I improved immensely under his care. I was even able to return to work part time for a few more years.

I am now on Hysone and although the adrenal insufficiency has worsened over the years, I find it much more manageable being on this medication.

Member Story 3

I have had several hospital admissions due to Addison's Disease over 40+ years.

In my case Addison's crises onset very quickly, mostly without warning.

Within minutes I am unable to stand or walk unaided. I am unable to shower, dress or feed myself.

My hospital stays are a minimum of 7 days and can be as long as 30 days, often followed by a 4-6 week stay in a rehabilitation unit.

When I am discharged from the rehabilitation unit, I am able to feed and sometimes shower myself but lack the stamina needed to prepare meals for myself. My gait remains impeded for 12-18 months or even longer and I leave using a wheelchair or a wheelie walker. I am unable to walk any distance (sometimes no more than 30 metres) and I am unable to drive for a minimum amount of 3 months.

My mental clarity is compromised and I find it difficult to follow conversations, make decisions and maintain focus.

My appetite is severely affected and consequently I leave hospital feeling extremely weak and fatigued.

For me, Addison's Disease can be very disabling and I require long periods of care and assistance before I am able to function independently



Member Story 4

When I was admitted to a hospital in the northern suburbs of Sydney on 5/3/19, I did not realise the impact it would have on me, not just at the time, but even today.

After being tripped by a dog leash at a café on the Northern Beaches, I fell and had glass embedded in both hands and my right forearm. I became very unwell very quickly and friends administered 100ml Solu-Cortef.

NSW Ambulance attended and were completely unaware of Addison's disease or the protocol they are supposed to have in place. They did not understand why the injection was necessary, and went on to advise the deep gash in my forearm would not need surgery, would possibly be glued, and that I was OK to have a coffee.

They suggested I might prefer to have friends drive me to a northern beaches Critical Care as there would be extensive waits at the new Hospital.

On arrival at ***** Hospital, I was told surgery would definitely be necessary but not available there, an X-Ray was taken and, because of the poor reputation of the new Hospital, I was driven by family to my local hospital in the north of Sydney.

I suffer from a number of rare health issues, but my Addison's disease caused the most problems for both myself and the staff at this Hospital, who simply did not seem to know what it was.

My MedicAlert bracelet was not examined at any time, nor was there any attempt to telephone to check my medical information. Whilst I was conscious and coherent, it was not an issue, but had I been unconscious or confused, it would have been a serious concern.

I was eventually examined by an orthopaedic registrar, and had to explain what Addison's was, and why I had had the cortisol injection. I gave full details of my medications, advising what my base rate of medication was for my Addison's. At no time was any attempt made to contact a member of the endocrine team.

I was self-medicating from my own tablets which had been brought from home, as no-one at the hospital seemed to know what to do with me. I was nil by mouth from approx. 8.30am, in pain and stressed, so needed to have additional medication to avoid an Addisonian crisis.

After 5+ hours, I was finally admitted to await surgery and was subjected to genital and nasal swabs 3 times because paperwork was not sent, sent incorrectly and finally correctly. Distressing at the best of times, and even worse when stress was already aggravating my Addison's.

A gowned female started to pull open my gown in front of others to attach electrodes to my chest without introducing herself, asking permission, or explaining the purpose.

All of these, together with delays in surgery pushed my stress levels through the roof – which had a major negative impact on my Addison's,

At about 6.45pm, I was seen by the orthopaedics registrar on evening duty, who confirmed he was hoping to operate that day



At 9.15pm he advised surgery was finished for the day but I was scheduled for approximately 9.15am the following day.

He was shocked when I advised him my wounds had not been cleaned. I said I was concerned about being highly susceptible to infections due to my Addison's and Hypogammaglobulinemia (immune system insufficiency) despite having iv antibiotics at the hospital as the wounds were still 'dirty.'

The registrar immediately arranged for a 4l saline "rinse" ie it was pored over the largest wound in my right arm. Nothing was done to my hands or the laceration on my left forearm .

I had been left for over 14 hours before anyone cleaned the wounds with saline, and only then after I asked. Sepsis is a real risk, and my immune system cannot cope with infection. Subsequently, I developed a post-surgical infection.

As I am gluten free, there was also no food available for me either that night or post op the next day.

Post op, I was seen by the Junior Medical Officer who blithely told me he had spoken with my endocrinologist who had instructed him to change the type of my steroids and reduce them to less than half of my maintenance dose.

When I said he must have misunderstood my specialist, because recognised protocol is always to increase not decrease steroids during and immediately post trauma or stress, he insisted he was right.

I did not know he was calling my specialist and at no time was offered access to the endocrinology team at the hospital, nor apparently did the JMO consult them for clarification

At this point, I felt I was at serious risk, and said I would discharge myself if he persisted. Even then, he sent me home with paperwork saying the change had been made. Had I followed his instructions, I could have gone into a coma and died. At no point was any mention made of the "Reach" program which I now know the hospital has for patients who feel unsafe, as I clearly did.

Fortunately, I was conscious, aware of my conditions and medications, and able to speak up for myself. I shudder to think what the outcome would have been otherwise.

As it was, I was left with no confidence whatsoever in the doctor (who has since admitted he was confused but didn't seek clarification) or in this hospital, and was terrified at the prospect of being admitted for anything at all in the future.

I have since been diagnosed by my GP with stress related high blood pressure, despite being previously healthy, and am now on medication for this.

This stress was exacerbated by the attitude of the hospital when I wrote in to register my concerns, and which then led me to go one step further to the HCCC in an attempt to safeguard the wellbeing, not only of myself, but of other Addisonian patients who enter the doors of Hornsby Hospital.

What should have happened?

Triage – medical conditions and their relevant seriousness recognised and wounds cleaned with saline whilst awaiting surgery.

Correct history taken by JMO who then seeks consultation from endocrinology team



Appropriate scheduling of surgery taking into account risks of Addisonian crisis exacerbated by pain and stress, and infection, following consultation with a member of the hospital's endocrinology team.

Swabs taken correctly.

ECG technician introduces herself and purpose of test.

Awareness of need to increase corticosteroids for Addisonians

'Reach' program (Recognise, Engage, Act, Call and Help) initiated when I expressed concern for my wellbeing

Discharge papers completed correctly (and no unnecessary medications dispensed ie I was issued with the medication I had refused to switch to and handed it straight into a pharmacy for disposal)

OUTCOMES

NSW Ambulance

I contacted NSW Ambulance who basically said, after several weeks, the two paramedics had been spoken with and were conversant with Addison's disease – clearly not!

A Northern Suburbs Hospital

After receiving a very patronising letter from the Director of Medicine, Dr K, in May 2019, which essentially implied I had misunderstood, been confused by anaesthetic, or was just plain wrong, I wrote back reiterating the issues, and was finally persuaded by HCCC to attend mediation with the hospital.

On entering the room, I received an immediate apology from Dr K who said the letter was totally inappropriate, offensive and did not address the concerns raised.

He confessed he had been on leave for 3 months and been pressured into signing the letter on his return without really reading it as it had taken so long to reply.

This set the tone for what proved to be a fruitful meeting.

This apology was repeated in a letter subsequently received, outlining the steps the hospital is now taking to improve the situation for any Addisonian attending the hospital emergency department.

Dr K has undertaken to provide education to all junior medical officers regarding Addison's disease, and for this to be in the early part of the year rather than at the end, as at present.

It was identified that there were perceived barriers for staff to escalate concerns to senior medical staff. The hospital is educating its staff about the 'Speaking up for Safety Scheme' which encourages staff to seek help when faced by such issues.

It was acknowledged the JMO should have consulted the endocrine team at the hospital, and that this could have had very serious consequences if I had not been very aware of my medications and emergency action plan.

All staff are to be re-educated in regard to the Reach program and the importance of implementing it.



Dr K and the Director of Emergency Medicine are to work on educating staff regarding the importance of medical identity bracelets and the need to check them as a valuable and life saving device.

My case was reviewed at the hospital's Clinical Review meeting, for discussion on ways in which the hospital could better manage such situations in future.

I was asked by the patient liaison officer to provide an impact statement which was presented to doctors to enable them to see the long-term effects on me as a result of the mishandling of my admission.

I also provided up to date brochures etc from the Addison's Association which were to be read by the endocrine team and amalgamated into their literature on Addison's disease, and we have been invited to forward and updates to them in the future

Junior Medical Officer

The JMO finally admitted he had been confused and concerned about seeking advice from a senior doctor and did not understand the severity of the condition or it's treatment. The HCCC has now amended their findings that he had done everything according to guidelines to state he should have consulted the endocrine team.

He should also have escalated to a senior physician when I expressed my concerns.

SUMMARY

I complained in the first instance to prevent this situation being repeated for any other Addisonian.

Whilst lengthy, very stressful and time consuming, I believe this hospital now has a better understanding of Addison's disease and how to manage patients with this illness, which makes it a much safer place for we Addisonians

Update sent October 2022

You have permission to use my story.

After meeting with the chief of medicine at this hospital and others, they agreed to bring forward the training re Addison's to an earlier stage of the junior doctors' educational roster so the knowledge was available sooner in the rotation.

They also decided there was a need for all junior doctors/registers to have re-emphasised the needs to check with their seniors if they were in doubt about correct procedures for someone with Addison's.

I obviously don't know if they actually did this.

This is relevant to the best practice that might be expected from a hospital involved in an inquest (in this instance the Peter Domachuk case) and it was disturbing that the procedures were not being followed already by 2019.



Member Story 5

This is a copy of what I sent to the hospital after my surgery. Please let me know if you would like more information.

As an aside, when I had chemo, my oncologist, Professor G, had had only one other patient with Addisons before. He took a lot of notes on how I adjusted my medication and how I reacted to the chemo.

I had a Lap R) Hemicolectomy on Tuesday 11th Jan at approx. 1:30pm. Because I have Addisons Disease, my surgeon Mr R, asked me to be admitted at 11:00am 10th Jan, so that a team could discuss and be prepared for my surgery. I had spoken to my Endocrinologist in Cairns but he assured me that he did not need to make contact with my surgeon or the hospital, as you would have Endocrinologists to provide advice. As soon as I was admitted, I was placed on a liquid diet which meant I had fasted for 26hours prior to my procedure- not a good start for a person with Addisons. The assistant surgeon, M (he was replaced by L on Tuesday and was not part of my surgical team) saw me at 6pm on Monday, ordering blood tests and asking me about what my Sick Day Management Plan is and my normal doses of Hydrocortisone. I would feel that my SDMP is far different from major surgery. He did not seem adequately informed about Addisons. After my surgery, I wholeheartedly believe that, if Nurse S had not pushed for me to be taken to ICU, I would have had an Addisons Crisis. Because my blood pressure was in the low 80s, she called for the special team three times. Two doctors asked me questions about how I felt, was I dizzy. Because I had had 100mg Hydrocortisone during theatre and then again at 6pm, there was a reluctance to give more. Many people with Addisons do not know they are going into crisis, but rather family around them can see the changes. Certainly, when I'm trying to function with normal day to day activities, I know when I'm low. Asking me questions 10 hours after surgery was not effective. I was extremely drowsy and Addisons makes you numb, almost calm. Also, the sicker I am, the more colour I have, making me look better. Dr A, your Endocrinologist, and wonderful doctor, did not know I was in the hospital until 9am Wednesday 12th January. It is very disappointing that a major Hospital did not ensure that the necessary specialists were consulted, especially for a rare disorder. I am angry that the reason I was admitted a day early was not addressed at all. If you would like to contact me to discuss further, do so.

Hospital Response – Annex 1, submitted separately as the letter was makred Private and Confidential.



Member Story 6

I have Addison's disease. I was diagnosed at the age of 19 after suffering a severe adrenal crisis which almost cost me my life. In the days leading up to my diagnosis, I would present at the Emergency Room of a large teaching hospital in Melbourne and after receiving an IV drip I would be discharged. Several doctors and nurses who treated me in those days dismissed my symptoms as stress from university. When my parents brought me into the Emergency Room on the day I was diagnosed, I was fading in and out of consciousness. It was a very close call.

Since then, I have unfortunately had many negative experiences when I present at the Emergency room of various hospitals. The main issues are a lack of timely medical care to prevent an Adrenal Crisis or urgent treatment during a crisis.

2015 -

- I presented at the the hospital's Emergency Department in the beginning stages of an adrenal crisis and was not administered my emergency Solu-Cortef injection.
- Even worse, my emergency injection kit was taken from me by the emergency staff in the ED so I could not even administer it to myself. This is the equivalent of taking away someone's EpiPen or insulin.
- After the 2015 incident was investigated, I was assured that an alert would be put on my file so that emergency department staff will be aware of my Addisons Disease. Unfortunately, I had similar problems the next time I attended.

21st November 2019 -

- Called for an ambulance but was told it would take a long time because my situation wasn't considered urgent
- My husband drove me to the Hospital Emergency Department. Despite us asking several times, the Doctor in charge at ER did not page the endocrinology registrar because she believed there wasn't one on duty. This is not true – there is always a registrar on duty and should be contacted immediately when a patient with Addison's Disease presents at ER.
- The doctor at ER gave me 100mg of Solu-Cortef and transferred me to ICU. I asked to see the doctor there and requested that he page the endocrinologist on duty, as they are the experts in dealing with an Addisonian crisis. Again, he said there was no endocrinologist on duty and that he would contact one in the morning.
- I explained that I needed another dose of 100 mg of hydrocortisone, which is the recommended treatment during an Addisonian crisis (100mg every 6 hours until patient is stable). I was only given 50mg until the doctor returned after some time (unsure how long), and reported that he had managed to get in touch with the endocrinologist registrar and was instructed to give me another 50 mg dose (total of 100mg) and organised for me to be transferred to the ward as an in-patient.
- As a result of delayed medical care, I became so ill that I had to remain in hospital for 4 days and spent weeks recovering at home.



22 January 2020 -

- On Wednesday 22 January 2020 I arrived at the Emergency Department, assisted by my sister, seeking treatment for a red-back spider bite. The spider's species was confirmed by the builder who was working at my property that day and my sister who is a hospital pharmacist with a level 2 first aid certificate.
- Despite showing the Triage nurse on duty the letter prepared by the hospital's endocrinology department, clearly instructing Emergency Department staff to contact the Endocrinology Registrar or Endocrinologist on call for further advice, she advised us that she can't page anyone and that she would inform the doctor on duty about my Addison's disease. The same thing happened when I was eventually seen by the nurse treating me in the ER.
- It was over two hours after I had received the spider bite when I was finally seen by the toxicologist on duty.
- I had to self-administer the emergency Solu-Cortef injection and was at no point assessed by an endocrinologist.

30 January 2021

- I presented to the Emergency department at the Eye and Ear Hospital with swelling, pain and redness in eyes and ears.
- I informed the Triage nurse that I have Addison's Disease and am immune-compromised.
- Due to an admin error on the part of the hospital, I was not seen for 7 hours despite repeatedly presenting at the reception desk and informing the Triage nurses that my symptoms were worsening.
- The problem with Addison's Disease is that a simple infection can deteriorate very rapidly and trigger a life-threatening adrenal crisis. Any delay in receiving medical assistance can be life-threatening.

1 April 2022

Large, specialist Hospital in Melbourne

- I was scheduled for a day procedure at a large Women's Hospital in Melbourne. Prior to my scheduled surgery, I had provided medical documents to the hospital about Addison's Disease. On the day of the surgery, I spent over half an hour with the admissions nurse going through my medical history and talking in detail about my Addison's disease. The hospital and medical staff taking care of me had all been informed and were aware of my medical conditions.
- As the procedure is performed under general anaesthetic, I had to fast and was not allowed any water even to take my medications. As a result, my last dose of medication was at 6 am and I was not called in to surgery until almost 3.30 pm. That is over 9 hours which is far too long between medication doses.
- By 3 o'clock in the afternoon I was feeling lightheaded having trouble staying awake, starting to experience mental confusion and my muscles were shaking. These are familiar symptoms that I have had before an Addisonian crisis.



- I had to self-inject my emergency injection because the nurses could not do it and the doctors were busy in surgery.
- The hospital was aware of my life-threatening medical condition and should have scheduled my surgery earlier because fasting and 9 hours between medication doses put my body under strain and triggered an Addisonian crisis.
- My life was put unnecessarily at risk

29 May 2022

- I was being treated at another major Melbourne hospital emergency department for a gynaecological problem when I began to experience the early signs of an Addisonian crisis. The doctor looking after me thought I would receive better care at the teaching hospital and organized for a nurse to take me there
- As instructed by my Endocrinologist, I self-administered my emergency injection as soon as the symptoms started and was then taken to the teaching hospital
- The nurse from this hospital explained to the admissions nurse at the teaching ER that I was having an Addisonian crisis and handed over my medical notes as well as my medical document clearly explaining what needs to be done to treat a crisis
- I waited over 4 hours to be seen by a doctor only to be told that I did not 'look sick enough to be having an Addisonian crisis.'
- 3 hours later, I deteriorated rapidly and had to request the nurses to administer another dose of my emergency injection. My muscle tremors were too strong for me to safely do it myself.
- It had been 7 hours since my last dose and medical protocol for treating an Addisonian crisis is administering medication every 6 hours.
- The doctor once again told me that in his professional opinion, even though he is not an Endocrinologist, I was not having an Addisonian crisis.
- At that point I realised I was not being taken seriously, even though I have 15 years of daily lived experienced with this disease and have lived through several crises, so I discharged myself and returned to the first hospital emergency department where I was treated with more respect.

Member Story 7

I would like to contribute two stories of medical neglect that have impacted me negatively as person living with a Disability.

Context:

In 2015 I was diagnosed with premature ovarian failure, which set off several tests which revealed I had antibodies throughout all my endocrine organs. In 2016 I was diagnosed with late onset Type 1 diabetes and in early June 2018 I was diagnosed with Addison's disease (Primary Adrenal Insufficiency). Prior to these diagnoses I was a fit and healthy individual, having spent most of my life as an elite athlete representing Australia for sport and working professionally as a psychologist.

I want to report two events that have impacted me adversely because of neglect and poor treatment by health providers. The first event was a hospital admission to the Epworth Hospital (Freemasons) in May 2018 only days prior to the Addison's diagnosis. The second event was regarding my treatment in emergency triage on Friday 6th Nov 2020 prior to my hospital admission at the Austin Hospital on 7th November 2020.

Incident number 1 – Private Hospital, Melbourne Late May 2018

What Happened

I thought I had a virus that looked like the flu, it had stopped me in my tracks, low blood pressure, fever, runny nose, sore throat, and a deep grating cough, my body ached, I slept for days, and couldn't walk without assistance, I felt a strange pulse in my head that made me faint. My muscles became uncharacteristically weak. I had lost a significant amount of weight – I am 5 foot 8 inches tall and weighed 50 kg. When I saw my GP she said I needed to go to hospital and wanted to call an ambulance but it didn't feel like an emergency, so I opted to let my partner take me to the Epworth. I was admitted the same day to the hospital in East Melbourne.

I felt like I was deteriorating in terms of fatigue and muscle weakness and kept falling asleep involuntarily. I was finding it hard to talk and I couldn't get to the toilet and back to my bed without the support of the nurse to walk. I didn't know what was wrong with me, but I knew something wasn't right which is what I tried to tell my treating hospital doctor.

The doctor assumed that I hadn't made peace with my diabetes diagnosis, she said to me

"you just have to make a choice and get on with your life...you can't let diabetes own you...I have a friend that has type 1 diabetes like you and she just ran a marathon...she doesn't let it get her down, you need to be tougher."

I was too tired to explain to the doctor that I hadn't let my diabetes diagnosis hold me back, in the 2 years since the diagnosis I had completed my Masters degree including submitting my thesis and finishing my internship, I moved in with my partner, spent 7 weeks travelling Europe, had started up my own private consulting practice, and travelled to Japan the previous November and successfully completed my karate exam, being honoured with the rank of Sensei.



I knew something was wrong, but all I could manage to say to the doctor was that 'I didn't take any pleasure in being in 'a sick role' and that I don't like hospitals', I told her 'I was a proud woman and if I had to get help to get to the toilet and wipe my bum then something was wrong.'

But she didn't believe me. The doctor told me I was probably depressed and needed to 'get on with my life' and that she wouldn't let me out of hospital till I could prove that I could walk by myself.

I was too tired to cry properly, the tears streamed down my face with distress and frustration, I started to second guess my experience, I felt like the doctor was telling me I was exaggerating my symptoms or seeking attention, her tone was patronising and belittling. I figured there was no point staying in hospital if I wasn't going to be believed so the next day, I used all the strength I had and 'willed myself' out of the bed to walk a few steps down the corridor to prove to the doctor I could go home. I remember my partner saying to me that I didn't look well enough to leave the hospital, but I told him I had to get out, that the doctor wasn't helping me, so I wanted to come home.

The day I was discharged from the hospital I got a call from my private endocrinologist Dr P telling me that I had to quickly have an ACTH stimulation test. ACTH signals your adrenal glands to produce cortisol. This test measures the level of cortisol in your blood before and after an injection of synthetic ACTH to determine if had Addison's disease (Mayo clinic website 2022). My ACTH test confirmed that I had gone into adrenal failure, I had to see Dr P straight away and commence lifesaving steroid treatment which is now required multiple times a day to keep me alive.

I firmly believe that it was only the quick thinking of my endocrinologist that ensured my medical safety, primary adrenal failure is life threatening and if she had not have requested the ACTH test I may have died or suffered significant further medical complications as a result of a delayed diagnosis.

I understand that Addison's disease is a rare condition and often misdiagnosed. The problem in this instance is that the doctor at the hospital didn't listen to me to start with to even be curious about my symptoms. If she had have taken me seriously and done more tests or requested a secondary consult, I may have had a different outcome in terms of a quicker diagnosis and treatment and without the psychological trauma that has left me with a distrust of the medical profession and a fear that I won't be adequately looked after when I need medical help. I believe that the doctor's bias towards me put my life in jeopardy, she effectively discharged a patient who was on the verge or possibly already by then in organ failure.

Why haven't I reported it officially till now? I have been wanting to report this since it happened, but every time sit down to write it, I feel scared that I won't be taken seriously. I told my family and friends about the incident, and some months after the Addison's diagnosis I started seeing a Health Psychologist due to my psychological distress regarding my mistreatment and overall difficulty with my physical and psychological adjustments to now living with two life-threatening chronic conditions. Four years later and with much support from friends, family and my psychologist I am finally in a place where I can find my voice to share this experience in written form, even though I am still working on healing from the trauma.



Recommendations for change

That the doctor in question be sent for professional development training on understanding the dangers of unconscious bias in medicine in addition to any professional communication training that centres around client centred interviewing for assessment and empathy.

Incident number 2 Large Teaching Hospital in Melbourne 6th November 2020

What happened

My birthday was the 6th November and we were in lockdown, my husband thoughtfully ordered us a fancy dinner from providoor to be delivered to our house to celebrate. Unfortunately, I ended up with food poisoning which resulted in severe stomach cramps, diarrhoea, and vomiting. This is a risk for people living with Addison's disease as dehydration and not being able to keep my usual steroid doses down puts me at risk of adrenal crisis which is life-threatening. I understood according to my 'sick day management plan' that since I couldn't keep my steroids down that I needed to go to hospital for immediate treatment. Both my husband and I were clear with the paramedics that my purpose for going to hospital was that I needed IV fluids and a dose of Solu-Cortef (intramuscular injected steroids) to avoid an adrenal crisis since I couldn't keep my medication down.

Because we were in hard lockdown my husband couldn't go with me to advocate on my behalf, but I heard the paramedics give handover to triage and they were clear about my presenting need was to be given IV fluid and IV steroids asap. But when I was put into the emergency bay I was left there for hours, when the nurses checked-in on me I kept trying to say I need IV fluid and steroids for my Addison's that the food poisoning symptoms are bad but that's not the life-threatening bit. I think at the time I entered triage the time was close to or after midnight, and I asked where the endocrinology consultant was and didn't get an answer. I didn't get the steroids till something like 8:30 the next morning? By this stage I was feeling significantly unwell, dehydrated, muscle weakness, confusion, still experiencing stomach cramps and now muscle cramps in my legs.

I was eventually admitted to a ward. When I got there the nurses asked me if I wasn't just pregnant. I told them I was infertile due to premature ovarian failure which is a part of my rare autoimmune condition. I think I stayed in the ward for approximately 3 days before I was stable enough to be discharged. During my stay the next nurse on shift asked me if I was pregnant, I asked him to read my file where it states that my ovaries don't work, I can't be pregnant, and to make a note for the next nurse on shift to not ask me this as it is distressing to me since I am still grieving for my loss of motherhood. The next nurse on shift asked if I was pregnant.

I was also given white bread with jam for breakfast – even though my meal preference was marked as 'diabetic' I was given jam – it was only because I looked at the packet to see that it was normal jam and not sugar free that deterred me from eating it. When I asked the nurse how I could give feedback to the food service management team that giving jam and white bread to a person with type 1 diabetes is going to make my medical management harder whilst in hospital the nurse said they didn't know the feedback process. Before I was discharged, I found a brochure in my top draw for feedback and managed to write down my concerns.

Since I didn't get support or guidance to give feedback about the food menu planning, I doubted I was going to get support to request an investigation regarding my lack of medical attention in triage. Again, this reiterated for me that because I have a rare condition that people don't take my medical



presentations seriously, and this negligence puts my life at risk for something that could be well managed if I could receive the emergency steroid medication as per endocrinology protocols for adrenal crisis or prophylaxis for adrenal crisis in the event of severe diarrhoea, and vomiting.

Recommended Changes.

I want to know if a clinical risk incident was submitted for a near miss pertaining to my medical event. Leaving me for hours in triage without an endocrinology consult and consequent medical treatment in terms of administering steroids, put me at a potential life-threatening risk.

I want to know if a clinical risk incident was submitted then what reviews and corrections have they made to policy and procedure to ensure this type of missed step doesn't occur again? Was this a case of medical neglect in that a staff member breached protocol by not contacting endocrinology for consult? Or was this a matter of not having a clear policy on adrenal crisis management, a problem with procedure or other?

I recommend that food service management review their nutrition guidelines for 'diabetic' meals considering the needs of people living with 'type 1' diabetes in terms of providing food that is high in sugar and high glycaemic index that could unnecessarily complicate a patient's medical stability whilst under hospital care (I have had this issue multiple times in hospitals).

I recommend that if hospitals are going to put posters on the walls stating that they are patient-centred and our voice matters that all staff are clear on how to support patients to give feedback. What the processes are, where the forms are kept and how to submit them or who to talk to.

I recommend that hospital staff continue to strive for education and support for sensitive practice – to read a patient's file and try to refrain from making 'off the cuff' judgements and statements that a woman of childbearing age presenting with vomiting must be pregnant. Being asked if I am pregnant multiple times whilst grieving for my infertility was the least of my problems in this scenario but you can see the cumulative effect of this experience amplified an unnecessary psychological distress.

As a side note, I was recently admitted to the same major hospital COVID-19 ward for a week, and thankfully had better care this time round in terms of the hospital staff listening to the paramedics handover about the nature of my condition and actioning endocrinology being involved immediately, receiving the appropriate steroid treatment and in correspondence with my private endocrinologist. Nevertheless, I still think that the story I have shared warrants investigation so that policy, procedure, or staff error can be corrected for future patients.

Many thanks for hearing my story



Member Story 8

I am a 70 year old woman who recently had to call an ambulance as I was extremely unwell. I am writing this as I would not wish for anyone else, including myself, to ever have to go through a similar experience.

I have Addison's Disease/Adrenal Insufficiency, amongst other health issues and due to contracting Covid became very ill. I had collapsed and passed out twice when my husband called 000 requesting an ambulance for me. When the ambulance staff arrived I told them that I was Covid positive and had collapsed twice due to also having Addison's Disease and was having an Addisonian crisis. I wear a medical alert bracelet also which states what is required in an emergency. I had previously put together a "Medical Emergency" envelope with relevant information for these types of situations which I carry in my handbag. My husband gave this information to one of the ambulance staff, who opened the envelope and quickly flicked through the paperwork without reading it then discarded it onto the couch saying "I don't see a list of your medication here".

They asked me what medication I was taking, including asking me why I had been prescribed steroids and how often I took them (cortisone acetate and fludrocortisone) which I once again explained to them. During the time that they were at my house, I told them four times that I had Addison's disease as did my husband on several occasions. Neither of the ambulance staff asked me any questions relating to my Addison's disease at any time. They decided to transport me to hospital and administered IV fluids as I was very dehydrated. My husband and I were told by the ambulance staff that when I arrived at the hospital I would be given another bag of fluids and then would be able to go home later that evening as I would not be required to stay there. During being transported to the hospital I was not given any medication for my Addisonian crisis.

When we arrived at the ED, one of the ambulance staff advised the Triage nurse that I was covid positive but did not advise them of my Addisonian crisis. After some time, one of the ambulance staff told me that they were leaving and had asked an ambulance staff member from another team to "wheel me into Emergency when I was to be taken through". Due to the number of people who were also waiting to be taken into the ED, and the number of new arrivals, who were processed before me, it was quite some time before I was eventually taken through and seen by a doctor. At no time did any member of the ambulance or hospital staff check on my condition whilst I was waiting in the outer ED area. I was eventually taken through and seen by a doctor who immediately administered the emergency medication that I required. I was eventually admitted into the hospital for five days, after being diagnosed as having "an acute Addisonian crisis".

This situation was very upsetting, particularly as I had all of the documents that I had been told to have "on hand" if I was ever in an emergency situation and went into an Addisonian crisis, so that I could give this to ambulance/medical staff who would then know what the symptoms of an Addisonian crisis were if they had not heard of this condition before, and what emergency medication I needed to have administered to me immediately.

I do understand that this is a very stressful time for hospital, ambulance and medical staff, but an Addisonian crisis is a life threatening condition which needed to be taken for more seriously than it was. I didn't feel that my case was dealt with appropriately, and that I did not receive the support that I should have been given, possibly through a lack of knowledge on my condition.



Member Story 9

NSW AMBULANCE SERVICE

In February, 2021 I had an unfortunate experience with NSW Ambulance Service.

I have Addison's disease, a rare autoimmune condition. I take hydrocortisone tablets three times a day to manage this condition. It can quickly become life threatening, called an adrenal crisis, when vomiting, diarrhoea or trauma occurs.

I live alone so I rang for an ambulance at 4pm as I had food poisoning, meaning my medication could not be absorbed. NSW Ambulance has a protocol for treating adrenal crisis, called the M24 Protocol. However, the call centre operator who, I was later told, is not medically trained did not listen when I tried to explain the reason why I needed urgent assistance. The paramedics finally arrived 2 hours after my initial call. By this time, I was in a very bad condition with increasing adrenal crisis symptoms. The paramedics had a good understanding of what was needed to stabilise my condition.

After a stay in hospital, I followed up with NSW Ambulance and was told, by letter, that "at the time of your calls (sic) there was an increase in demand for emergency ambulance resources in your area." I live in a busy suburb of Sydney in the Hills District, not in a rural region.

I tried to follow up this letter on a few occasions but was unable to contact the Executive Director, Clinical Operations, NSW Ambulance, as advised in the letter. The two concerns I wanted to raise were the long wait time of 2 hours in a life-threatening emergency and how could I be flagged in the system so this does not happen again if I need an ambulance?

It took me 3 months, with many emails and phone calls, to finally reach a resolution to my situation. I was actually told that, if I needed to call an ambulance again, to say I have chest pains or breathing difficulties and this will get me a faster response. As a truthful person who does not like to "queue jump" I would be most reluctant to take this advice unless it was actually happening.

The eventual outcome was that I am now "red flagged" in the NSW Ambulance system. I had to give permission, due to privacy concerns, for my medical condition to be displayed in their system, which I was happy to do.

Unfortunately, I feel I can no longer rely on, or have trust in, the NSW Ambulance Service. I have had to make alternative arrangements with my family and GP to get emergency care should I need it again. This has also impacted my mental health, with frequent flashbacks, and this causes me anxiety.

CHANGES THAT SHOULD BE IMPLEMENTED

- I understand that call centre operators cannot have knowledge of all medical conditions. However, people should be given the option of providing their medical information to



ambulance services if a delay could become life-threatening with the worst outcome being death.

- I would also like all state and territory ambulance services to have a uniform and consistent protocol for dealing with adrenal crisis. The patient should be treated promptly as too much hydrocortisone given will not cause harm but, if it is not given, death can occur.
- I am also mindful of the time and energy it took for me to reach the “red flag” outcome. There would be many other people who, for whatever reason but possibly health related, could not persevere to achieve the outcome that I did. It should be easier to reach the “red flag” outcome.

Member Story 10

On 25th April 2021, I had a spontaneous episode with extreme pain in my upper and lower back region and continuous vomiting. An ambulance was called and I was transported to a hospital in Qld and then overnight by flying doctor to a larger hospital. First to the surgical unit and then to Urology. CT scans were performed but no MRI. I was there for two and a half days before the pain subsided, with no definite diagnosis. I was discharged unable to walk or stand. They were caring but very busy and understaffed.

A week after discharge I was sent by the urologist to have another CT. The referring doctor did not call me with results for several weeks. My local GP received the results also and passed on the findings. I was diagnosed with a spontaneous bilateral adrenal gland haemorrhage. No treatment was recommended except they had sent me home with soda capsules. Weak, skinny and exhausted.

Long story short, on 15th September 2022 a local doctor was relieving at the clinic I go to and immediately felt that there was a need to pursue the reason for my still weak condition. The doctor contacted an Endocrinologist for advice and from there it went quickly. The doctor sent me for a Synacthen test (ACTH) and I was diagnosed with Addison's Disease and received hydrocortisone. **Seventeen months** after my haemorrhage. Quite a lot of personal stress and worry during those months. Including a vaginal prolapse in July 2021 and a full hysterectomy on 5th September 2021.

No cause has yet been found for my adrenal crisis. Since 17th September 2022 I am seeing a visiting Endocrinologist. My condition is now improving.

In conclusion, I feel that within the general practice and regional hospital community there is insufficient knowledge or training for recognising Addison's Disease or Adrenal Insufficiency as it is quite a rare occurrence. I think some follow up would have allowed my Addison's to be diagnosed much earlier. I am grateful that I experienced no serious sickness or injury in the 17-month period without medication.



Member Story 11

Classic Symptoms of Addison's Disease – in retrospect!

My story starts over 30 years ago and I have resisted putting it into writing – maybe because the days and months leading up to my diagnosis were the most harrowing of my life?

I was a married woman of 39 at the time. We had 4 children, all at school, and my husband and I were both teachers, and also were involved in community work. I had ceased having my periods many years before, at age 27.

A year before my diagnosis in 1988, we took two other children, along with our family to the Brisbane Expo on a camping trip. I found it really hard work with 6 kids in tow. Shortly after that trip I was driving a school bus one afternoon in November and felt absolutely overwhelmed with tiredness. I pulled over, drank water and had a break. I safely completed the journey, thank goodness.

In the afternoons on my way home from school I would stop at a corner store to buy packets of salted chips, and devour them. At home I made myself salted lemons, and would eat them. I had never done this before. I often could not eat my breakfast as I felt nauseous. In the morning when I woke up I had to hold on to the bedpost as the entire room was swimming around my head! Boxing Day I could hardly get out of bed, and the tiredness and nausea were getting worse. I was dragging myself around, and had to crawl up the stairs in our house sometimes. I lost weight and was turning browner. During January I went to the GP several times, and he did tests, no significant findings? I took the test results one night in total desperation to the ED of a major teaching hospital. The triage said I could not be seen by a doctor in ED as I was seeking a second opinion! I went to the staff physician at my workplace. She listened to my story but did not have any suggestions.

I went to another GP. She listened to my symptoms (tiredness must have been top of my list?) and she told me to go home and go to bed for a week. Several friends commented on how well my lovely bronze tan was holding on after my Queensland holiday. (Addison's disease has a nickname of "bronze skin disease" I was to discover later!).

I went to yet another GP. This woman listened intently and wrote notes. She said she wanted to call a Physician and book me in to see him as soon as possible, which turned out to be 5 days later. The physician asked a lot of questions, including questions about the health of my parents, and advised me to come to see him for tests at the hospital in 3 days' time.

I started vomiting during the night and could barely stand up. I was sure I was dying. My husband drove me to the hospital and helped me into the Emergency Department. They put me in a wheelchair and I was "flopping" out of it.

I was taken into the treatment area without much waiting time, I was injected with life-saving hydrocortisone. In 10 minutes, I felt better than I had done in months! What a miracle drug this is!



I still had to stay in hospital for 5 days and was quite weak on returning home. I had a whole term off school as I took a long time to recover. Later I would realise how close I had come to being “diagnosed on the slab”.

I eventually returned to work (teaching) but it took me a while to adjust and accept that I now lived with a chronic condition- for the rest of my life. Once I embraced this fact, I was able to get on with living every day as a gift. I willingly take my Cortisone Acetate(Cortate) and Florinef every day. I have made myself available to help train endocrinology students ever since but it is the recent moves to introduce awareness of rare diseases into GP and intern training that I regard as most valuable. After all, once I got to the specialist Physician, diagnosis of Addison’s disease followed immediately. It was the GPs who failed to recognise that further expertise was needed when a tricky case come up.

What I know now! November 2022.

My Symptoms of Primary Adrenal Insufficiency in retrospect:

- Craving salt
- Darkening of my skin
- Low blood pressure indicated by dizziness getting out of bed in the morning.
- Loss of appetite and nausea
- Weight loss and muscle atrophy
- Tiredness to the point of utter exhaustion.
- No menstruation
- In final stages, vomiting. probably confusion

My advice to people wondering if they have adrenal insufficiency is to be persistent. Keep going back to the doctor. Follow up.... don’t get complacent.

Member Story 12

An accidental diagnosis

In 1973 I was in an Adelaide hospital ward, with salmonella, and (the official description) “a wasting disease”. I was 24 years old, and I knew I was dying.

A month before, I had consulted my GP because of concerns about deteriorating energy levels, and a tendency to collapse if I stood up suddenly from a squatting position. When I got home from work, I would occasionally feel the need to immediately lie down on the floor. I was working as a pay clerk at the time, and work performance was fine. I asked the GP if I could have a blood test to look for abnormalities, and mentioned to him that I was planning on joining a group of friends for a journey on the Birdsville Track. The test results had come back by departure time, but I was told that only the Doctor could discuss them with me, in a formal consultation. I concluded that they couldn't be serious, so left the next day for Leigh Creek in my Kombi Van, together with my mates from the Off-Road Vehicles Club, in various four-wheel drives.

Next morning at Leigh Creek, I decided to turn back. I felt that I would be endangering not only myself but my partner and her two boys (and our Lhasa Apso dog) who were accompanying me, if I continued. Heavy rain that had started at Leigh Creek continued, and I persevered with driving through the day. I was getting weaker, however. My partner was unable to cope with the heavy mud we were encountering. We reached Port Broughton successfully, and spent the night at the caravan park.

Next morning, I got to the Men's toilet but didn't return ... I was too weak. The dog came looking for me, and had a peek under the door. It's not usual to see a Lhasa Apso's eyes but I remember them opening wide with concern when he saw me!

By the next morning I was in hospital, isolated with reverse barrier nursing due to the salmonella which would have got a mention as a Notifiable Disease in the paperwork sent to the lackadaisical GP I had consulted (but never again). Due to good hygiene, my travelling companions had not acquired the infection.

Time passed, punctuated only by the never-ending sound of the iron lung operating in the only other occupied room in my wing, and visits from family.

It became apparent that the available medical staff were unable to diagnose my underlying problem. I had concluded that I was probably going to die.

Unexpectedly, I had a visit from a Tamil doctor who was interested in the medical facility from a professional perspective. He examined my notes, asked me a few questions (including, with a trace of a smile, “Do you have Indian blood?”), and left the room for a brief discussion with the local endocrinologist. Not long afterwards, the endocrinologist re-appeared to announce smugly that the mysterious “wasting disease” had been identified, and could be treated.

There was never any admission that my survival was due to the unnamed Tamil doctor. Replacement therapy for my Addison's Disease began with Cortisone Acetate, and Florinef. My health recovered steadily, and has been good since. The results of a fairly recent survey done in Norway indicate that so far, I have survived



over 9 years past the average age of death of male Addisonians. All thanks to the unnamed Tamil doctor!



Member Story 13

My Story of Medical Neglect and Eventual Diagnosis

The Beginning

I was born a 26-week-old female in 1967 with 14 major things wrong. Looking back now, it seems my adrenal glands never worked properly in the first place. I also had Diabetes, so neither did my pancreas.

I had a liver failure and pancreatitis when I was eight, and heart failure at 12; no explanation. I think these diseases affected everything in my body. Six months off school and yellow as a banana, doctors thought my school had an outbreak of Hepatitis A; but the test came back negative and I was the only one in the school that had it. For as long as I could remember I would throw up morning, noon and night, I had no temperature control, my blood pressure was that low that most doctors thought I would be dead by morning, and terrible pains in the legs if I fasted. I would be that sick and screaming in pain that my parents would have to get the night doctor to come to our house. He told my parents it was growing pains. I couldn't keep the weight on. Then I would come good for a couple of days. I would eat like a horse, feel terribly ill and throw up again. It was just my normal... My family couldn't take me anywhere without me being sick. So, my diet consisted of basic foods, I never ate any form of sugar, it would make me sick, I often had a fetish for lemons, raw eggs and of course salt. I would crave it. I had a tan when I was younger, but that was just attributed to the Australian climate. It couldn't be Addison's Disease... it was too rare. The first 12 years of my life were filled with every childhood disease imaginable; multiplied by ten. My immune system was shot.

1980s

I caught the Epstein Barr Virus in Grade 8. I was that sick I missed the last 2 months of school and all my exams. That was the end of my vaguely well childhood. I was never the same after that. A blur of doctors, anxiety, terrible pain after exercising, feeling gravely ill, tea coloured urine, weight loss and severe fatigue. I got my driver's licence in 1985 but could only drive to the shopping centre. I would be sick and then basically pass out on the floor. My mother ~~would have to drag me to the café, give~~ gave me copious amounts of coffee and chips and gravy loaded with salt. Again, I was told it's just post viral fatigue. I felt like I was dying. Often, I don't remember how I even drove home.

1990s

Now came the "Yuppy Flu" era. I kept getting sicker and sicker to the point of not being able to get out of bed to go to work. I was skin and bone and could hardly keep anything down. The fatigue and stomach cramps were overwhelming. There were many days when I didn't even know what day it was or even cared. Most days I lay curled up in my bed unable to move. Using what energy I had, I dragged myself to every doctor imaginable to try and find out what was causing me to be so sick. I saw Physicians, Immunologists and Cardiologists. 90% of my wages were spent on doctors, medicines and supplements that would presumably work and give me my life back, but never did.

My GP referred me to a hospital in Brisbane for more testing. At this point I was living in genteel medical poverty. While my friends were falling in love and getting married and going on



honeymoons, I was paying doctors, getting admitted to hospital and having test after test done. My friends and I were now worlds apart. Specialist after specialist could find nothing that even resembled 'Addison's'. The problem was that the doctors weren't listening to me, they just kept on doing the same tests over and over again. A simple ACTH Stimulation test could've diagnosed me in an hour. But I was refused.

Instead I was diagnosed with anxiety, cardiomyopathy of unknown cause, a vague myopathy in my legs, Cerebral Palsy and an extremely fatty liver.

Doctors told me that Addison's Disease was only a paragraph in the textbook that took 20 minutes to go over in class time, it was too rare to be followed up. They told me to go home, and stamped my medical records with Chronic Fatigue Syndrome and closed my file. Back home, I remember being that sick and sitting in the doctors office with a blood sugar of 2.5 and barely conscious. He handed me a bag of lollies. Took my Blood Pressure which was 60/40, looked at me and said, 'You must be really fit... goodbye'... and I was shown the door.

I managed to hold down a job at the local school as a Teacher Aide and Admin. Assistant for twenty years. I was that sick that most of the time my principals kept telling me to go home. I just sat there in my chair, I dreaded having to do playground duty, sports days or teacher aide training, as it nearly killed me.

I was told to report to the doctors office at least once a month, but the same old test were performed. I was labelled a nuisance by the doctors. I overheard doctors label me as too complicated and to steer clear of me. I had exhausted my choice of doctors in Country Queensland and Brisbane.

I was at a dead end. By this time I had been in my room for 23 years. I missed out on everything: having friends my own age, independence, graduating, relationships, marriage and children. I had to watch everyone that I knew meet someone and get married, while I was either hooked up to an IV or simply left in my bed to rot. I had become an invisible nuisance. No friends or relatives cared enough to see if I was ok, or visit. My social circle was reduced to zero. I only had my dogs. My family were sick of me being sick. I had become a medical liability. So I took an overdose of Panadol, which I happened to survive, because later I found out I couldn't absorb tablets. There were no mental health awareness programs back then. It was one of the darkest periods of my life.

The Year 2000

By this time I had spent most of my life in bed. I was going into deep coma like sleeps for days, and then I would wake up. My Doctors still put it down to chronic fatigue syndrome, until one day I went into anaphylactic shock and my GP put me on prednisone. It was the first time that I felt nearly well in 40 years. I still only weighed 42kg, by this time my parents told me to give up work before I died there. The Cerebral Palsy and Muscular Dystrophy qualified me for the disability pension, so at least I wasn't out on the street.

By this time we had computers and a thing called the Internet. I kept on coming back to stories about people with Addison's who felt better once they were hydrated and had salt and I said to my mum, that's me. I'm 40 years old and I weigh 42kg. My GP did a strange thing, he referred me to a very good Physician named Dr N. By this time I had an Endocrinologist and was diagnosed with Type 1 diabetes and then Type 2 because of the steroids. Then I had Cushing's because I had too many



steroids, as I needed the prednisone to control the anaphylactic shock. I also had an immunologist now called Dr K. He too had no idea why this was happening and why I felt so well on prednisone. IVIG was prescribed for my immune system and the myopathy. It was wonderful. For the first time I had energy, I could stand up, I wasn't throwing up. And then they decided to wean me down off the prednisone and that's when the problems started again.

This time I was having actual withdrawals from lack of Prednisone, so the doctors tried me with Solu-Cortef, it stopped it for a while, but when it wore off, I would be moaning and writhing in bed, again, I couldn't keep still, it was like having withdrawals from heroine. Again, I felt like I was dying, this would start happening when I got down to 20 mg of Prednisone. By this time I was labelled a nuisance, a person who liked being in hospital because I had nothing better to do, a whinger, and an attention seeking patient who needed to be placed in a psychiatric ward. Still only the same tests were performed over and over again, when I mentioned Addison's I was told to be quiet and that I should stop googling everything on the internet.

The days I nearly died:

2017

It was my brother that saved my life that day. And of course my Endocrinologist who didn't have many privileges in the hospital; so his hands were tied. I just happened to get admitted through the cardiology ward because I had heart failure, I was in that much withdrawal and distress again that I couldn't keep still. Previously, my brother called the ambulance, they were great, but said they had seen nothing like it and that they were not allowed to give me Solu-Cortef; it wasn't an approved drug.

By the time they got me to the hospital, I was hysterical, having small seizures and was half paralysed. I told the nurses and the doctor repeatedly that I needed Solu-Cortef or I was going to die. I was repeatedly ignored. My Physician Dr N was on holidays, so a younger upcoming physician was reluctantly put on my case. He told me to pull myself together, it was all in my head and that I had an addiction to prednisone. My Endocrinologist, walked in and saw me, but because he didn't have any privileges at the hospital, he was disregarded. He told my brother that I needed 50mg of prednisone to pull me out of the dive in order to save my life. He couldn't tell the covering doctor this because the doctor on my case was a higher authority... so basically he was just letting me die.

I learnt early on how to save myself. I always carried an extra bottle of prednisone in my bag for emergencies. So I took 50mg without telling him and the change in an hour was miraculous. I could move my arms and legs, no withdrawals and my speech and the droop in my face had improved. I made the mistake of telling the doctor the next morning what I had done and he went totally ballistic, screamed at me and told me how dare I self-medicate without his authority. By this time I had had enough and I screamed back at him and told him that I had to be the live patient. I had to save myself, because he wasn't going to do it and I was dammed if I was going to let myself die. Then he mumbled under his breath, 'Well I suppose I should've given you some prednisone'. I was disgusted. You could clearly see the difference in me, I was talking and coherent.

But before I could count to 10, this well-dressed lady in a size 8 suit came in with her clipboard. One of many psychiatrists I would see. You could tell that she was young and that her brain had been removed from her head, she couldn't think outside the box as to what had happened. She told me that my GP of 37 years had agreed with her that I had an addiction to Prednisone and that I needed to be evaluated by a psychiatrist and be in the psych ward where I could slowly get help for my addiction. I was absolutely devastated. I thought that my GP knew me better than that.... He had



betrayed me. I had no words any more. I just wanted to die. By this time I had been treated badly by my family, friends, every doctor I had seen and now my GP of 37 years.

I was discharged with a warning and a threat of being put in the psych ward if I came back. When I went back to my GP, I kept my mouth shut, but I was warned by him not to upset any of the upcoming physicians in the hospital as they were going to replace my doctor when he retired. If I wanted to be treated by them in the future I had better tow the line. I had no words. As for my mental health, I felt like everything was closing in on me and that suicide was probably my only option. It was only the love of my dogs that saved me.

2018

By 2018 I was having more serious withdrawals from lack of Prednisone and was being rushed to hospital once a fortnight. Still no mention of Addison's Disease on my medical records. After being admitted again with the same withdrawal symptoms, a Chinese lady doctor that was covering for my doctor that day, walked in and said, 'I've seen this before, you've got Addison's Disease and you're having withdrawals because you haven't got enough Prednisone in your body', I was absolutely stunned, it was the first time that any doctor had mentioned Addison's Disease. She put me on an IV with saline and gave me 100mg of Solu-Cortef. Within half an hour my body had stopped wreathing in pain, I stopped vomiting and I felt like I could breathe again. I'm sure that if this doctor had not been on call that day, that I would've died that night. Although I was told my blood pressure was within normal limits, I felt my body was shutting down.

Again, she said she couldn't say much as she was only covering for my doctor, nothing was mentioned about Addison's Disease in my chart. Again, I don't know if she was real, or if she was an angel, but she saved my life that night. The other doctors were still telling me that I couldn't have Cushing's disease and Addison's Disease and Diabetes in a couple of years; it was unheard-of.

2019

The withdrawals were getting more and more frequent, it seemed that my body just couldn't absorb tablets any more. It didn't matter how much Prednisone I took, and I took the least possible dose, because A. I didn't want to get fat, and B. it was wrecking my bones and my teeth. Again, I ended up in the hospital on the weekend when my physician was away. I always got nervous when this happened.

Unfortunately, a new doctor was left to care for nearly 70 patients in one night, which was absolutely ridiculous. I could see that this was not going to end well. I could feel my body shutting down, but he just waltzed in, quite annoyed, and told me that my blood pressure was within normal limits and to stop whingeing. There were student nurses on at that time and I remember just breaking down and crying telling them that I needed prednisone or I was going to die. I told the nurses this as well, they agreed, but their hands were tied and even though they told the doctor I needed some, he dismissed them.

By this time I was absolutely stressed out of my brain and I had read somewhere that sometimes if you take a hot shower or sit in a hot bath that it stimulates the adrenal glands and my body might make just enough cortisol to save me... I was wrong. By this time they had taken all my medication off me and made sure that I couldn't get the prednisone I needed. I was diagnosed with a psychiatric



problem called Conversion Disorder; Neurological symptoms such as paralysis or withdrawals from a drug that cannot be explained by the doctor. I was being watched closely.

So half an hour later I ended up in the shower chair with the shower going, I manage to turn it off before I blacked out. Here I was with a blood sugar of 1.5 in a coma under the shower. It was just as well that I was sitting in the shower chair because if I had collapsed standing with the water running, I probably would've drowned and had a head injury. A nurse found me unconscious an hour later apparently, and the MET button was activated.

I vaguely remember being dragged from the shower onto the bed with oxygen. I was apparently in a coma I had low sugar but normal BP. It really rattled the nurses and the student nurses, as I had just predicted a few hours before what would happen, they saw the whole thing unfold in front of their eyes, they thought that I had died. I was in a coma. It was only after IV glucose and hydrocortisone was given, that I briefly opened my eyes. The nurses when they went off duty apparently rang the hospital to see how I was doing. They were visibly shaken. I spent a few hours in ICU and then was taken back to the ward.

The next morning my Physician was on and found out what happened and he couldn't understand why I collapsed in the shower... Unbelievable. He told the other doctor, who now was sweating bullets, that he had done the right thing. There was no apology to me. I really thought this time that it was the end. I was basically on my ninth life.

At last, to clear up any confusion, my physician decided at last to do a ACTH Stimulation Test, to prove once and for all if I had Addison's Disease or not. Guess what, my cortisol was so low it hardly registered on the test. Again, no apology from anyone, but I did get a saline drip and all the Solu-Cortef I needed. I was never questioned again, and Addison's Disease was at last written down in my chart with a proper diagnosis and proof that I did indeed have Addison's. I was too wrung out to cheer, just hurt and very sad that I nearly died three times before anything was done. I was determined never to feel this bad again.

Covid and The Way Forward 2020

When Covid hit in 2020 and everyone was having a nervous breakdown because they were being isolated, I was not perturbed by this at all, as my life had been lived in isolation, rejection and medical negligence. I had lots of time to scour the internet for advances in treatment for Addison's Disease and I came across an article where they had put Solu-Cortef in an insulin pump and programmed it to the circadian rhythm of your body. This 'new' therapy had been trialled in the US and UK for nearly a decade. After reading numerous peer reviewed articles, the evidence stated that every candidate that used the pump for Hydrocortisone Therapy for Addison's, noted a marked improvement in the control of their disease, energy, feeling of well-being and a smoother delivery of hydrocortisone which enabled them to live their best life. Again, I was gobsmacked. But this time in a good way!

I was still having absorption issues with my tablets and was being rushed to hospital once or twice a week. Addison's was not the question now, control of this disease was my problem. I didn't have any, not with tablets anyway. I just couldn't control my Addison's anymore. Like my Diabetes before the pump, it was taking over my life and leaving no room for anything except doctors, hospitals and



medication, I couldn't live like this anymore. I had nothing to lose. Medically and emotionally I was at rock bottom and could go no further.

So I got in touch with a professor at a hospital in London and his secretary gave me the name of a doctor in Brisbane who had started pump therapy on his Paediatric patients with Congenital Adrenal Hyperplasia, basically kids born with adrenal glands that didn't work properly. The pump was working well for these kids, again stats showed 100% improvement in each patient's control of the disease and quality of life.

I had nothing to lose, so I made an appointment to see my endocrinologist and I talked to him about pump therapy... he readily dismissed it and said that there was no proof that it changed the outcome of people living with Addison's disease. I begged to differ. I was on an insulin pump for my diabetes and it had changed my life in three days, it was much better than injections with an insulin pen which I couldn't also readily absorb.

I just happened to have an extra insulin pump as the company had changed hands and had given us a new pump to make the transition easier. This Endocrinologist came to my ward every day and calibrated my old pump with Hydrocortisone until she got it exactly right. This doctor was my miracle. The pump took a few days to kick in, but again it changed my life. It's like I didn't even have Addison's, anymore.



Member Story 14

Please find below some bullet points about my diagnosis journey and treatment since being diagnosed with AI.

Symptoms first became so severe that I couldn't work in March 2017. I had confusion, brain fog, headaches, high heart rate, nausea and diarrhoea, trouble sitting for long periods and a lot of headaches etc. I handed in my notice at my full time job when it became too dangerous to drive (near misses on the way to work) and I didn't feel safe anymore. My workplace also refused my request to work shorter hours (GP note requested this).

2017 - 2019 Path to Diagnosis (Diagnosed by Crisis 28 Nov 2019 after Neurosurgery)

- **Number of GP appointments - 45** (Out of pocket cost \$1,262.45) - 4 different GPs (two main, two ad-hoc)
- **Number of Specialist appointments - 65** (from Medicare statements) (Out of pocket costs \$5,275.50)
- **Number of Diagnoses - 21:**
- **Number of hospital admissions/procedures without steroid cover - 8**
 - Two presentations to ED's in 2018, where they just sent me home and offered me diazepam. I presented thinking I'd had a stroke, because I couldn't talk properly, I had wobbly footing, and I couldn't remember names etc. They did take me straight through but after I saw a Neurologist registrar they sent me home. I began to think it was pointless going to hospital as they never believed me and I only came out sicker (due to the expended energy).

The first time during this 'hell period' that adrenals were mentioned to me was in consultation with the Cardiologist and I mentioned my small appetite, big thirst and my salt cravings (because he suggested increasing salt intake and fluids to treat POTS) in April 2019. My morning cortisol at that time was 328 nmol/L with a lab ref range of (150-535nmol/L). In May he wrote to my GP and said I should do a Synacthen Stimulation Test (SST). I had this on 18 June 2019. I remember having to lay the whole time waiting for the test and feeling really unwell. In August 2019 I have a consult letter from the Cardiologist stating "She had a SST on 18 June. The baseline cortisol level was lowish at 220 nmol/L, but still within the normal range. There was a suboptimal cortisol response to synacthen administration, with a 50 minute sample yielding a cortisol level of just 390nmol/L. About half an hour after that, the level was only 480 nmol/L." He goes on to say "the patient had already started fludrocortisone at time of testing. Fludrocortisone can interfere with the assay and therefore I will discuss these results with a biochemist or endocrinologist colleague. We might need to repeat it after a time off fludrocortisone".

The next time I heard from the Cardiologist was a voicemail dated 11 October 2019, followed by a letter sent to my GP. He suggested repeat testing and said he would send the forms, but I was flying to London the next day on the way to Spain for my urgent Neurosurgery. My GP emailed on 18 October and suggested I show the letter to the Spanish team and I did. They provided steroid cover for my surgery. About 2/3 weeks after the surgery, I was weaning down the steroid dose so I could be formally tested for Mast Cell Activation Syndrome (they diagnosed me by questionnaire and symptoms seen during my hospital stay) and got down to about 10mg hydrocortisone when I



became extremely unwell with vomiting, body pain, confusion, inability to speak. I was then admitted by ambulance back into hospital for 5 days.

They discharged me with a reducing hydrocortisone regimen and said to see an Endocrinologist in my country of residence.

2020-2021 Recovery from Neurosurgery and Addison's Crisis/ COVID isolation

I was unfit to fly back to Australia, so got urgently referred to an NHS Endo (Endocrinologist A) in London (I just had to keep calling for cancellations). She looked at me in my wheelchair and neck brace and said "you don't have addisons" but she sent me for tests that confirmed I did. Once she had the blood test results, she treated me much differently and during the course of the year in the UK, between her and my GP, we used day curve tests to ensure I was on the right maintenance dose of hydrocortisone.

Return to Perth on New Years Eve 2021

Went into hotel quarantine. Unable to get a GP appointment until Feb 2020 - showed him letters from Endocrinologist A and he wrote a referral in the public health system to Endocrinologist X. He wrote a referral for cortisol tests (day curve) at a Perth hospital - I had to make a number of calls to explain what I needed.

As I was due to see multiple specialists and GP was unsure of the root cause of my problems (we thought either Addisons, MCAS or Gastro) I ordered a 'Pain & Symptoms Tracker' and started logging my food, meds, physical symptoms, sleep, activity and fluid intake. I began logging this on 20 October 2021. During this time I was trying to do distance study (10hrs a week), walk daily (1hr a day), and read. I was on a dose of between 62mg and 86mg but averaging 72mg. I was having a lot of loose bowel movements, pain, fatigue, and stomach cramps after eating - my quality of life was not great.

- **2 Dec 2021** Endocrinologist B emailed to advise that the pharmacogenomic study showed I didn't have a problem metabolising hydrocortisone. I told him I was trialling his new dose but that my husband had walked out and asked if I should double the dose (168mg). He replied and said "stress dosing will depend on how psychologically distressed you are but I suspect a couple of days won't hurt given your news". My husband was still in contact and coming around and causing distress (we had an anniversary weekend away booked 10-13 Dec and we had booked Christmas lunch together. I was hoping we could resolve our issues as he was hyperfocused on work projects and probably burned out.
- **15 December 2021** - I reached out to my GP by email because he knew the history of mine and my husband's relationship and my medical complexities. I explained that I was socially isolated and didn't have the support I needed in place. I included the below dosing schedule (table below), GP forwarded this onto Endocrinologist B who replied "you might benefit from social work input if you have no other support. I would suggest trying to taper the hydrocortisone dose somewhat after 5 days or so. Please be mindful of glucocorticoid excess such as weight gain, thin skin, easy bruising". ***There was no mention of the risk of steroid induced psychosis.***
- **20 - 22 Dec 2021** Patient called own ambulance

I was extremely anxious and sleep deprived (sleeping only 2hrs 30 mins on night of 19 Dec). I had extreme fatigue and confusion and fell asleep from c.4pm for the rest of the day/night, slept through



dinner and doses and (according to Fitbit data) woke up around 9:15am the next morning. On waking I felt terrible, and unable to get up. I crawled to the bathroom and proceeded to vomit numerous times. After some time I crawled back to my phone, tried to stand and fainted. I got through to the triple 0 operator but had trouble speaking and answering her questions. I eventually managed to say "Addison's Crisis" and tell her I had a flag on my address with an Ambulance. I don't know how long I was on the phone to her but I asked her to instruct the crew that arrived to administer my injection as I'd heard about problems relating to this. She was good and helped me with my breathing but the whole thing was scary as I didn't feel I had long left (especially without the injection). I actually begged her not to leave me as I didn't want to die alone. I was kneeling on the floor with my head between my arms trying to not panic (as I knew that I would use more cortisol), I managed to crawl to the door and open it. *When the ambulance finally arrived they just hung up my mobile and weren't told by the operator to inject me.* I have instructions that I have hEDS and a fused neck but they made me get upright myself and this injured my neck and shoulder quite severely. On being upright, my footing was poor and I fell on my front steps and injured my hip and left side. Once in the ambulance the paramedic accused me of trying to suicide as I'd mentioned my husband had recently left. I was having trouble communicating due to confusion and I showed my wallet cards on adrenal insufficiency, hEDS and got out my emergency injection that they'd collected from my house. They refused to inject me and wouldn't let me inject myself. I believe this was due to having high blood pressure. I did not receive IV hydrocortisone until after I was triaged. I think this was around 2 hours after my emergency call. The discharge report states that I had Hypokalaemia and Adrenal Insufficiency. I had taken my "Pain and Symptoms tracker" with me so they could see my medical info. I was unable to contact Endocrinologist B, despite trying numerous ways. I was assigned to Endocrinologist C who included in the discharge notes the following:

- "Usually on total daily dose of hydrocortisone 60 - 80 mg Has been stress dosing to total daily dose of 160 - 190mg of hydrocortisone for 3 - 4 weeks due to social stressors and factors
- Had her last hydrocortisone dose at 4pm on Monday, and fell asleep for 9 - 12 hours, before waking up on Tuesday with vomiting and having forgotten to take her PO tablets
- As patient was unable to swallow, patient presented to ED
 - //Diagnoses//
 - 1. Primary adrenal insufficiency
 - Known to Endocrinologist B privately, previous investigations RE aetiology; negative autoantibodies - no evidence of Addison's disease
 - 2. Ehlers Danlos syndrome
 - Complicated by cervical spine fracture and had C-spine stabilisation surgery
 - 3. Postural Orthostatic Tachycardia Syndrome
 - Previously seen by Cardiologist A
 - On Fludrocortisone 100 mcg
 - //Observation and Examination//
 - Hemodynamically stable, afebrile
 - Saturating well on room air
- 1. Hypokalaemia
 - K 2.1 on admission, likely due to huge doses of hydrocortisone steroids for a long period of time, and a hard wean (accidently) over 16 hours, leading to inadequate cover
 - Resolved with IV supplementation, K 3.7 during admission
 - Given 100mg IV hydrocortisone STAT in ED, and then 50mg TDS for 24 hours, and then

stepped down to PO hydrocortisone as per the weaning dose below

2. Adrenal insufficiency

- Given the complexity of patient's issues, usual physiological dose of hydrocortisone of 20mg is insufficient enough to maintain her day to day activities presently
- A complex weaning regimen was provided, to wean from 150mg hydrocortisone to 100mg hydrocortisone total daily dose on 22/12, then 90mg total daily dose on 23/12, and then 80mg total daily dose on 24/12, to about 60mg total daily dose, with the aim to be between 60 - 70mg of total daily dose of hydrocortisone upon seeing Endocrinologist B in the outpatients private setting
- Recommended total daily dose when physically well would be 20mg hydrocortisone for baseline physiological requirements, only increasing doses for physical illness and higher steroid requirement in this context, not for emotional stressors. We recommend a slow continued wean in the community to reach this target [please see below regarding the weaning regimen]
- No signs of steroid excess i.e. Cushing's on examination, however ongoing risk at high doses if not weaned

3. Social issues

- Social work input regarding increase in services and help at home was provided during the hospital admission
- pt declined psychology input this admission

//Plan//

1. Please continue the weaning regimen of hydrocortisone as provided by ourselves and by the pharmacist for the next few days, until you make an appointment with Endocrinologist B. The aim is to get to your stable basal dose of total daily dose of 60 - 80mg hydrocortisone Please note, the preference is to go down to 20mg hydrocortisone as recommended for physiological requirements, with stress dosing for physical illness only, as discussed during the hospital admission.

2. GP Review in 2 - 4 weeks time regarding the above and any other support services required"

When I saw the social services member of staff in the ward she said I didn't need her services, I complained but received no further help. They did provide a shower chair for use at home. I got home by taxi where I was left to look after myself. My neighbour met me, helped me carry in the shower chair and I went to bed. I was in a lot of pain from the damage of the fall and I felt traumatised by my treatment. I was exasperated that there wasn't more help available for me.

I followed the above guidelines, but did have to updose (4mg) when I attended the GP on 23 and again on 24 Dec (this was to fill in NDIS forms).

I believe this rapid taper caused me confusion issues (along with the social stressor of my husband walking out and me not yet being independent) and I began being unable to log in my "Pain & Symptoms Tracker".

The period between Christmas and New Year involved visits in my home from my ex and his brother. It is a very fuzzy time and they did not log my medications. I was trying to organise independence (bank account, centrelink, food, social care), I was trying to do this without the knowledge of my ex. As I had been ill for such a long period, I had no one outside of my ex and his family that I could contact for regular support.



- **31 Dec 2021 Neighbour called ambulance**

During these hospital visits, my bag containing my emergency injections and my steroids were removed from me. I did not receive my medications on time, and when I explained how important this was, the nurses just said they were busy. I felt frustrated but hoped I could manage at home until my booked appointment with Endocrinologist B on 19 January. I lost both my emergency Injection Kits in hospital (labelled with my name, adrenal insufficiency etc). Once again, I was not provided with social assistance at home. I was using an office chair on wheels to get from my bedroom to the kitchen to get drinks/ food (when I stood I felt faint and had a very high heart rate), I was also still suffering injuries from the ambulance and fall.

- **8-16 Jan 2022 Admitted to ED by separated husband/brother in law**

This time, I was taken to hospital by my brother-in-law and the difference is that the principal diagnosis became "confusion" with comorbidity of "Hypokalemia" - I was admitted to the Neurology ward because I had suffered a number of falls while at home trying to care for myself. I have no record of my meds for this time (25 Dec onwards) but I think my Mum via video call from the UK and my ex were trying to check up on me taking my doses. I was not sleeping or eating well, was suffering diarrhea and vomiting and I was only having varying times of clarity. I needed support to control my addisons. When I seemed confused to my ex, he instructed me by Whatsapp to take 20mg of hydrocortisone which I would have done as I trusted that he knew best (as he was not confused and had attended all of my consultations with me).

During admission, my meds were taken off of me and locked away, as were my replaced emergency injections (that I did not receive back). I did not get my doses on time and I was subjected to many invasive tests that were not required. I was suffering flashbacks to previous hospital visits where there was pain or where I had to remove my clothes (vulnerable). I was suffering from paranoia and didn't know who I could trust to help me. I think this is quite valid as I had no one who understood my condition (to advocate for me) and had been abandoned by my husband. I could also not get answers from the doctors as to why this was happening and how they could help me.

The following Invasive investigations happened to me during this admission (looking for "organic cause of confusion" despite the fact that Addison's IS an organic cause for confusion), I believe this to be because I was unable to advocate for myself as my brother in law said I wasn't acting normal:

- Lumbar Puncture (very painful)
- CT Brain x 2 (one on admission for confusion and one following a fall on the ward)
- MR Head
- CT Chest and Abdomen
- Whole body CT
- Pelvic Ultrasound with reasoning of confusion (I was menstruating at the time and I remember refusing to do it but the summary says they had patient consent)
- I tried to escape the hospital twice and believe they called security (I had bruises and pain following them restraining me)
- Trying to leave was seen as "unusual behaviour" and they put me on form 1A.
- This led to a psychiatric review who suggested an EEG and a one on one nurse special. Following this, form 1A was revoked as no evidence of psychiatric illness was found.



- Reviewed by Occupational Therapy, Social Work and Physiotherapy who felt the patient had significant cognitive impairment
- No cause found for patient's current presentation but patient discharged against medical advice.

- **18 Jan - 31 Mar 2022**

Ex-husband returned me to hospital, principal diagnosis became "psychosis" with comorbidities of "addison's disease, ehlers danlos syndrome, mast cell disorder, postural orthostatic tachycardia, hypokalaemia, hypophosphatemia", there was no one to care for me at home, I was admitted into a "locked ward" because I required "a high degree of nursing care", they commenced me on antipsychotics which made me erratic and very fidgety - the opposite of what I am usually like with addisons. This was a scary time where I had no access to my meds and was reliant on the nurses giving them to me on time. I tried to explain how important this was but on numerous occasions they sent nurses to inform me that I had received the wrong dose. I believe I needed gradual tapering and consistent levels of hydrocortisone (probably at a stress dose due to emotional stress) and no antipsychotics in order to regain my health and independence. With the different conditions I have, I required help to ensure I had longer term care in place at home (since my husband left and refused to help). The discharge report says that the OT's did submit an NDIS request that further information would be required once I returned from the UK. I was only allowed to be discharged because my ex agreed to accompany me to the UK to be cared for by my mum. I had to spend two additional weeks in hospital because my ex was "too busy". They discharged me with a 7 week weaning schedule down to 20mg a day. I was not feeling at all well at this dose.

- **UK Consults/ Day Curve Test 2022**

On return to the UK, I contacted Endocrinologist A and explained what had happened and that I was suffering anxiety, PTSD and trauma following my separation and my medical treatment. I explained that the doctors had put me on a weaning schedule but that I was not doing well at all on it. Endocrinologist A organised a day curve test to ensure I had adequate cortisol levels in my blood.

- **Endocrinologist B Consult Aug 2022**

I was hoping Endocrinologist B was going to be able to explain to me what had happened as I did not see him during my prolonged hospital stay and I believe we only conversed via email. When I told him I was following his advice of 80mg dose and that I had asked him at the consult Nov 2021 specifically if I would double that for stress dosing he had said yes. He said that his notes had me on 52mg. I hope to show Endocrinologist B his notes in my tracker at my next appointment so he can see that I was following doctors orders.

Member Story 15

Event 1: Northern Beaches Sydney 2020

My sister was in a diabetic low and not conscious. NSW Ambulance was called. On arrival I informed the ambulance officers that my sister was an Addison patient who needed to have cortisol administer straight away to avoid her from going into a Addison crisis. They informed me that they could not administer the cortisol, but that I could do it. I asked them if they could prepare the syringe for me which they were willing to do. Luckily for me my sister become conscious and I was able to administer the cortisol orally.

Event 2: Nambour Qld 2022

My sister was in a diabetic coma and non responsive after what was a diabetic low. The ambulance service was called and told that she was a type 1 Diabetic and had Addison which required the administration of cortisol through injection. The lead ambulance officer indicated that they did not had any cortisol with them and that they could not administer the injectable form that I hand in my hand. They were however, willing to prepare the syringe and allow me to administer it to my sister. Again luckily my sister become conscious and I was able to administer the cortisol orally.



Member Story 16

On 25th April 2021, I had a spontaneous episode with extreme pain in my upper and lower back region and continuous vomiting. An ambulance was called and I was transported to Emerald Hospital and then overnight by flying doctor to Rockhampton Base Hospital. First to the surgical unit and then to Urology. CT scans were performed but no MRI. I was there for two and a half days before the pain subsided, with no definite diagnosis. I was discharged unable to walk or stand.. They were caring but very busy and understaffed.

A week after discharge I was sent by the Rockhampton urologist to have another CT in Emerald. The referring doctor did not call me with results for several weeks. My local GP received the results also and passed on the findings. I was diagnosed with a spontaneous bilateral adrenal gland haemorrhage. No treatment was recommended except they had sent me home with soda capsules. Weak, skinny and exhausted.

Long story short, on 15th September 2022 a local Rolleston doctor was relieving at the clinic I go to and immediately felt that there was a need to pursue the reason for my still weak condition. The doctor contacted an Endocrinologist for advice and from there it went quickly. The doctor sent me for a Synacthen test (ACTH) and I was diagnosed with Addison's Disease and received hydrocortisone. **Seventeen months** after my haemorrhage. Quite a lot of personal stress and worry during those months. Including a vaginal prolapse in July 2021 and a full hysterectomy on 5th September 2021.

No cause has yet been found for my adrenal crisis. Since 17th September 2022 I am seeing a visiting Endocrinologist in Emerald. My condition is now improving.

In conclusion, I feel that within the general practice and regional hospital community there is insufficient knowledge or training for recognising Addison's Disease or Adrenal Insufficiency as it is quite a rare occurrence. I think some follow up would have allowed my Addison's to be diagnosed much earlier. I am grateful that I experienced no serious sickness or injury in the 17 month period without medication.

From that day on I haven't had one withdrawal, I stopped vomiting and gradually got my life back. There will always be medical issues that I will have to deal with all my life. I guess the saying, 'never give up', seems cliché. However, there are a few things you need in order to not give up. You need an Endocrinologist that believes in you; that's in your corner. You need a family or an advocate that believes in you, someone that will protect you when you are so sick that you can't even make sense. And a great support group. This is really an overview of what happened to me during my life just with Addison's, I haven't discussed the other 13 medical conditions, but that's for another day and another chapter of my life.



Member Story 17

Outcomes of two contrasting experiences in hospital

Firstly I must say that like a lot of people with Addison's Disease or Adrenal Insufficiency I have a very complicated medical history with many interwoven medical conditions which can trigger and impact on each other. Over the last 5 years I have had more Adrenal Crisis than I can count and am well known to my local private hospital emergency department.

The plus side is that I have had many experiences with ambulance paramedics, nurses, doctors and hospitals. I have had absolutely horrendous treatment and been told it's all in my head and my numbers are all fine nothings wrong with me through to wonderful experiences of medical professionals who actually ask my opinion and work together with me for the best outcomes. This has given me experience and knowledge of how my body responds and what works best for me. It has enabled me to advocate for myself and what I need and to be able to refer medical staff to my medical letters, action plans and best practice treatment. This doesn't mean it easy by any means. I must also say that I am one of those rare people who has a hydrocortisone pump as I have poor absorption of oral medication due to Crohn's disease.

Here is my example of both good and bad experiences during one medical emergency and it occurred at the same hospital.

GOOD EXPERIENCE

I was out for dinner with a group of business friends when despite all planning and preparation my dinner was cross contaminated with something I'm severely allergic to and I had an anaphylactic reaction. I have the most amazing team of specialists who look after me and I have detailed management plans and so I followed the plan.

The venue called for an ambulance stating anaphylaxis and Addison's disease and the paramedics arrived within 10 minutes lights and sirens. By this time I had taken antihistamines, antiemetics, epipen and intramuscular emergency hydrocortisone injection. In me an anaphylactic reaction will trigger an Adrenal Crisis.

After a brief assessment and upon finding out that I had been in ICU previously for anaphylaxis the paramedic called for MICA and administered more adrenaline. Adrenaline triggers angina in me and so things were just deteriorating. The MICA arrived and after an assessment got me on a stretcher and into the ambulance where I was given more adrenaline and a drip was inserted, pain killers given and fluids connected.

We then headed for the closest hospital (public not private) where I went straight into the resus bay and was promptly attended to by a lot of people, all who had a job to do. They were wonderful, caring, supportive and most importantly collaborative, I was part of the decision making process and we developed a good plan that everyone had agreed on and I quickly improved.

TURNED TO BAD EXPERIENCE

After about 3 hours I was moved to short stay ward and the doctors had a shift change. This is when it all started to fall apart. The IV analgesics were wearing off and the oral ones hadn't kicked in yet so I had chest pain related to the adrenaline I had needed. The hydrocortisone had started wearing off and I was nauseated and dry retching. My back and abdomen were burning, it's a pain that's hard to



describe, sharp and burning, I only experience it during a crisis. My head was foggy and I was beyond exhausted.

I was stuck in a loop that I couldn't get out of, the low steroid levels were contributing to intense pain which was then a stressor on my body burning up the hydrocortisone which was causing symptoms that were stressing my body. At this point I'm almost begging the nurse for another dose of IV hydrocortisone and after that some pain killers. In that order because at least the pain was keeping my blood pressure up.

For the next 3 hours *I kept asking the nurses to help me and get the doctor*, do something. At the same time I was using my pump to give myself hydrocortisone bolus just to keep from passing out. I asked for the doctor and was told she's caught up with a sick patient, I asked to see the consultant on call and was advised this is the public system at 3am there's no one here but residents. I asked them to call my private endocrinologist which they refused to do. I asked for the doctor in charge of the ED, it was the doctor assigned to me.

So I waited another 2 hours and endured 2 more ECG's and more blood being taken which made my IV access stop working and I just resigned myself to the fact I was going to end up in ICU again as I couldn't get the message across that this was serious and I was too exhausted and basically gave up.

The doctor eventually turned up and tried to placate me with statements like she's in charge and there's a lot of sick people here (my brain says and what I'm not sick???), my numbers aren't that bad (again little voice in my head says no thanks to you that's only because I have a pump and basically kept my system going).

Then I got the clincher, the statement that told me she really didn't understand Addison's disease/Adrenal Insufficiency at all, she said the nurses kept calling me but I didn't think that you needed hydrocortisone or pain killers because you were doing so well when we sent you to short stay, I thought you would have a nap and then go home at 6am (again brain says ahh hello that's because I had been given hydrocortisone and pain killers and they were working - thank goodness I'm the only one that can hear the little voice in my head because by this point I was totally frustrated and just fed up)

Why did she not communicate this with the nurses who would have told me and I could have addressed the issue 5 hours earlier and I probably could have gone home at 6am. Instead I had a nurse in each arm trying to find a vein to insert an IV to give me the medication the doctor now realised that I desperately needed. I also spent about 8-12 hours longer in emergency & short stay. This clearly demonstrates that one person with a different opinion, one person who has never met you and doesn't know your history, one person who thinks they know better than specialists who have treated you for years, one person who chooses not to follow documented medical management plans can derail best practice medical care which is clearly working.

REFLECTION

I don't know what the solution is but this shouldn't be allowed to happen. Maybe it's not just one thing but a combination of things that are needed. Things like national clinical guidelines, more education around rare conditions, a program around working with patients and their families to get the best outcomes or instructions on following management plans written by specialists or even better practices around handover and continuity of care.

It's hard enough to manage on a good day when you have multiple medical conditions that impact your life and when you're having a bad day it's almost impossible. Constantly having to advocate for yourself and fight to have people hear you and help you is truly exhausting especially when you're already not coping well.



Unfortunately this is a condition where things can change dramatically very quickly and as a patient sometimes it's just too hard and takes more energy that you simply don't have, so you give up and resign yourself to the fact that your going to get a lot sicker before anyone will take you seriously and actually do something.

It's an invisible illness and there is a lack of understanding around when you look bad and your numbers are bad it's too late. Treatment needs to start while things may still "look" good because if not things are going to turn bad and people can and have died.

Common sense is anything but common and very difficult to teach but somehow the message needs to get across to medical professionals that quite often the patient knows the best practice treatment they need and that working with the patient rather than dictating to the patient will bring about the best outcomes.



Attachment 1. Invitation to submit a story

Disability Royal Commission: Have you a story to share?

A number of members have already contributed stories of neglect that some people experience while living with Addison's disease/adrenal insufficiency for our submission for the Disability Royal Commission.

Many people live normal lives with Addison's disease/adrenal insufficiency once they are diagnosed and adequately medicated. Others struggle on a daily basis, and this can be even more problematic when people have other conditions. Often people experience temporary disability prior to diagnosis, and some people have horrific diagnosis stories.

As an Association we hope that contributing our stories will raise the Commission's awareness of temporary and permanent periods of disability that can arise because of being misunderstood or neglected, or unreasonable assumptions made by the medical profession.

If you too would like to share your story of experiencing disability while living with Addison's Disease, please send your stories to bronwyn@addisons.org.au.

We will collate the stories and send them as a single submission to the Commission. Please send your stories before 1st October, 2022. You will not be identified personally in the joint submission. If you would like to make your own submission to the Commission, you are most welcome. You may wish to do this if you want your case listed in the report.

Website for the Royal Commission into Violence, Abuse, Neglect and Exploitation of People with Disability:

<https://disability.royalcommission.gov.au/share-your-story/make-your-submission>



██████████r 2022

Private & Confidential

Mr. ██████████

Dear Mr. ██████████

Thank you for your feedback in January this year in relation to your experience at ██████████ when you were admitted on 1 ██████████, 2022 for ██████████ surgery.

I would like to sincerely apologise for the delay in formally responding to your feedback. The response was delayed due to staff and medical specialists being on leave and unable to respond to a range of questions we raised. I thank you for your patience.

I am very sorry that we did not meet your care expectations in relation to appropriately managing your Addison's disease. While we aim to provide the best care and experience to our patients, we are disappointed that we fell short in this instance.

██████████ is committed to investigating patient feedback. In order to provide this response, I reviewed the care we provided and contacted the various specialists who were involved in your care including the Intensive Care Unit (ICU), Dr ██████████, Dr ██████████ and Dr ██████████ (endocrinologist). The specialists each came to review your medical record and provided a response to the issues that were raised.

As you note in your letter, you have Addison's disease and I ██████████ arranged for you to be admitted the day before surgery so the team could discuss and prepare for your surgery. Dr ██████████ assessed your case before surgery and charted a course of hydrocortisone, but did not seek advice from the endocrinology team.

Dr ██████████ provided care during the surgery and immediately post-operatively. She relied on Dr ██████████'s management plan, and again did not involve the endocrinology team.

After your surgery you developed a range of signs and symptoms consistent with an evolving Addison's crisis and were transferred to ICU for ongoing management. At that time Dr ██████████ our head of endocrinology, was consulted and your endocrinology issues were expertly addressed.

Your experience and feedback highlighted the importance of patients with Addison's disease being reviewed by an endocrinologist as part of the pre-operative work up. As a result, the endocrinology team have reinforced to the surgical specialists the need to initiate a routine review by an endocrinologist of any patient with Addison's disease, prior to surgery.

I appreciate the time you have taken to provide us with your feedback and concerns, which we have taken seriously. We were able to use a number of your observations to review our processes and provide feedback to our staff and visiting specialists. I trust that this review will ensure our processes have changed to provide better care for future patients with Addison's disease.

We thank you again for providing feedback and contributing to [REDACTED] commitment to providing high quality and compassionate care to our patients.

We wish you well and hope this response addresses the concerns you raised.

Yours sincerely,

A large rectangular area of the document is redacted with a solid grey box, obscuring the signature and any accompanying text or title.