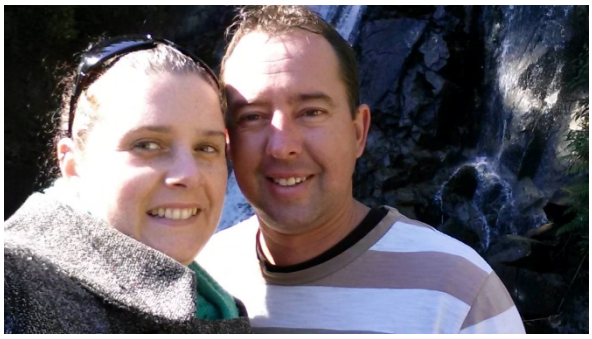
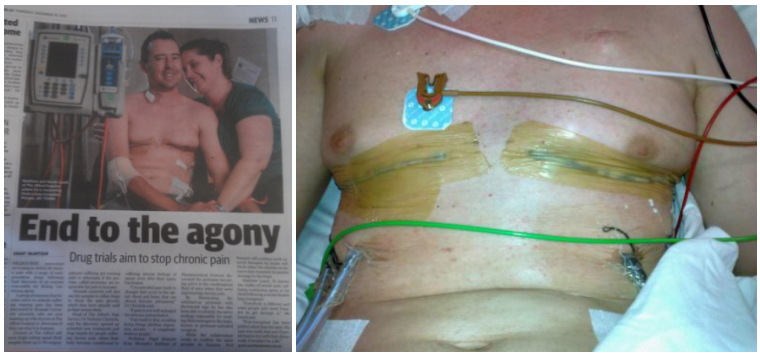
Nicole’s story



My husband Matt & I at Steavenson Falls in Marysville.

I was an otherwise healthy 37 year old. I am the carer of my husband Matt, who has Cystic Fibrosis. He essentially looks after himself now that he's had a double lung transplant and is now very healthy. I work full-time as a Research Assistant at Melbourne University, researching the damage that occurs to the bladder after a Spinal cord injury.



Matt & I in the Herald Sun (left) after Matt’s double lung transplant December 2013.

We live in Geelong and I commute from Geelong to Melbourne for work. I am also a very keen netballer, who had just returned back to playing netball after a knee reconstruction in February 2014. I also love to bake, especially for Matt, and he enjoys eating whatever I make.

In February of 2015 I was feeling sinusy and stuffed up. I usually get that way every now and then. This persisted and I finally went to the doctor after trying pretty much every over the counter treatment (I mostly went to their ‘After hours clinic’, as I could get there after work, instead of taking time off during the day for an appointment, and it was usually a waiting time of at least a week to see my regular GP). This Doc told me that I had a sinus infection and gave me a double dose of antibiotics, which eventually cleared up the infection. It was now June. At the end of the month I started getting pressure in my ears and put up with it for a while, but it didn't go away. Just before we went away on holidays in August, I called into the doctors again and was given more antibiotics for a middle ear infection - I’d never had a middle ear infection before.

Matt & I on Hamilton Island. Still having middle ear issues.

I had troubles with my ears equalising during our holiday, and avoided swimming. We'd driven from Geelong to the

Whitsundays with Matt and my in-laws. Some of the mountains along the way nearly had me curled up on the floor in pain as my ears just wouldn't pop.

I went back to the doctors after we came back and was prescribed a different lot of antibiotics for my middle ear troubles. Halfway through the course I woke up with incredible pain in my feet. It was horrible and I had trouble walking, so back to the doctors I hobbled with this new problem. This doctor (not my own GP) diagnosed me with an 'Inflammatory Arthritis'. It took me ages to get going in the morning, and the 'Arthritis' started progressing to my ankles, knees and fingers. I was having difficulty playing netball, but struggled through games in Goal Shooter, often in a whole lot of pain. I was also having difficulty at work with my fingers being so sore and swollen. It was difficult to operate a pipette at times, but my determination got me through the pain. The doctor also diagnosed me with some anxiety as I was having some pain in the middle of my chest and shortness of breath episodes. He prescribed me some CBT therapy and referred me to a Rheumatologist for the feet issues.

The shortest Goal Keeper ever (Left). Another netball injury kept me out of the winning grand final game (right).

These symptoms persisted, but I continued to put up with them. In November this doctor ran some blood tests, which showed that my inflammatory cells were high, I was put on prednisolone. In December I started feeling like crap. I was feeling nauseous and lethargic. A couple of weeks before Christmas I was off my food, this was very unlike me, as I love food.

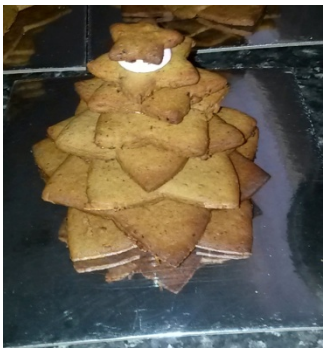

That's the biggest sticky date pudding in Parkville.

The week of Christmas I developed a dry cough and more breathlessness. I called into the doctor's clinic again with my new symptoms. He didn't think that they were significant, probably my anxiety, but said that he'd send me for a lung function test and a chest x-ray. I was told it would probably not find anything significant in either test. I phoned both places to see if they were open after Christmas, and seeing they were, pencilled them in on our calendar to do them the first working day after the break - the University closes over the Christmas break, so I had the time off. The doc gave me some medication to help with my anxiety. He said it had a lead-in time and that I had to take a low dose for a while before going onto the full does. He said that it would make me feel like crap, and gave me his personal mobile phone number to call him over the break if I needed anything.

I took the first dose of the anxiety med on the morning of Christmas Eve. I was finishing off the decorations on some baked Christmas presents for the extended family. I felt terrible, I was nauseous and had rolling bouts of dry retching. I sat a bucket next to me whilst I was trying to pipe the buttercream and place on small decorations. We

saw my parents and brother and sister-in-law that night. I was still nauseous and dry retching, but also peeing a lot, even though I wasn't drinking much.

On Christmas Day we went to my in-law's place for lunch. I felt even worse than I already did. I barely touched my Christmas lunch. I laid on my mother-in-law's spare bed and dry retched all afternoon, in between waves of nausea. We had Christmas dinner at my parent's place, where I was there in body only. I ended up vomiting up my dinner, this just wasn't right.



Some of my baked Christmas gifts. The biscuit tree was what I was trying to decorate whilst I was sick.

I called the doctor on his mobile on Boxing Day morning about the medication and how it was making me feel, he said I could skip a couple of days and start it again after Christmas, so I didn't take it Boxing Day. I didn't feel as bad as I did the days before, but I still felt terrible. We fronted up to my husband's extended family BBQ. I still barely touched a thing.

The night after Boxing Day I noticed that I was coughing up blood. At some unGodly hour I was coughing, a lot of blood came up, and it was coming out of my mouth and nose. At that stage I thought I was going to die... There was something seriously wrong with me! I nearly woke Matt up to call an ambulance, but I thought I was having a panic attack, so I remembered the CBT I was using and managed to convince myself that I was ok and tried to get back to sleep.

The Monday, still a holiday, I caught up with a friend for a coffee. I was weak, and I seriously don't know how I drove to the shopping centre to meet her. I only drank a soft drink, whilst she had brunch and a coffee. I was still coughing up blood and was very weak. I was coughing so much that it was making me incontinent. I would pee myself every time I coughed, I couldn't control it.

The next day I begged Matt to take me into Geelong for the lung function test and chest x-ray. Even though it is only a 100m walk between both places, Matt had to drop me out the front of both places as I was so weak and my feet were so sore. I struggled through the lung function test. It took me 3 goes to get the initial reading, then another 3 goes after ventolin to get the second reading. I coughed up blood in between, but hid it from the techs.

Matt then took me for my chest x-ray. The technician was quite concerned and called in the radiologist. He was also concerned and called us into his office. He showed us my x-ray, it was very white – similar to what Matt's lungs looked like before his transplant. He called the GP I was dealing with, but he wasn't at the clinic that day. The radiologist made the decision that he'd write a letter for me and sent me across the road to the emergency department of Geelong Hospital.

With the letter and my coughing up blood, I was seen in the ED pretty quickly. They took some blood and transferred me to the RAP unit, as they thought that I had pneumonia. Pretty soon after I was transferred to the RAPU, I was seen by the renal registrar. She told me that I was in renal failure and that I was being transferred to ICU.

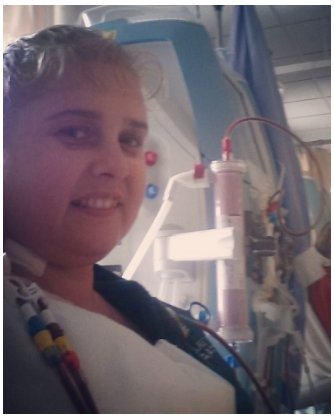
Everything went very quickly from then on and I have no concept of time over the first couple of days. I was put on lots of IVs (methyl-pred and cyclophosphamide) and given oxygen through positive pressure nasal prongs. I had a

central line inserted and they started plasmapheresis that night. The first few days I had plasmpheresis in the morning and 'filters' in the afternoon. The 'filters' are a portable dialysis-like machine that they could use in ICU. I also had a kidney biopsy and a small subcutaneous implant inserted in my abdomen (Zolodex) to help prevent damage to my ovaries by the Cyclophosphamide. My bloods showed that I had an eGFR (a measure of kidney function) of 4! I was also positive for Anti Neutrophil Cytoplasmic Antibody (ANCA)-Proteinase 3 (PR3) and Glomeruli Basement Membrane (GBM) antibody in my blood. My kidney biopsy also showed that I was positive for ANCA, and that of 34 glomeruli counted (glomeruli are the filtering components in the kidney), 30 has crescents in them (scarring). I was given the diagnosis of having both Wegner's Granulmatosis (WG, cANCA-PR3 positive) and Goodpasture's Syndrome (GPA, GBM positive). I was visited by my now nephrologist, who gave me the whole picture, balls and all. She told me not to be surprised if my kidneys don't recover from this and to prepare for more permanent dialysis.

In ICU at Geelong Hospital.

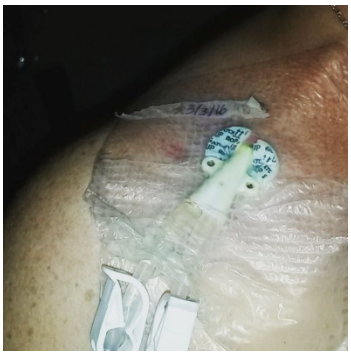
I spent 8 days in ICU, before being transferred to the ward. I was now dialysing 3 days a week, with plasmaphersis every second day. I had a semi-permanent vas cath inserted in ICU before I was transferred to the ward. I was improving, but was still quite weak. I was still getting out of breath doing the little things, like having a shower and getting dressed. I was weaning myself off the oxygen too.

I was in hospital for a total of 16 days. I finished plasmapheresis on the day I was discharged. The 'Inflammatory Arthritis' had disappeared, but I'd developed DVTs in both legs whilst in ICU. I was on daily Clexane injections together with EPO three times a fortnight to get my red blood cells up, as I was anaemic. I continued to dialyse three days a week and was getting stronger and stronger each day.



Dialysis selfie. I dialysed for only 8 weeks.

8 weeks after I was diagnosed with WG & GPA, my renal function had recovered sufficiently that I no longer required dialysis. I was monitored closely, as I could go backwards and require dialysis again, but that never happened. Two weeks later I had my vas cath removed.



Bye bye vas cath. Semi-permanent line for dialysing.

I was told that if any of my serious symptoms came, like coughing up blood, I was to go straight to the ED again and get them to call the renal registrar. One night, about eight weeks after my discharge, I coughed up a blood clot. I woke Matt and we discussed whether or not this could be a once off event or the start of something. We erred on the side of caution, as I had since started coughing up small amounts of blood, and at 3.30am we hopped into the car and drove to the ED. We spoke to the triage nurse, and I specifically told her that I had WG and GPS and was currently a dialysis patient. She told me that she'd never heard of the conditions. She ignored our request to ring the renal registrar. About 90 mins in, Matt asked her why I hadn't been seen yet. She told him that I didn't look sick and apparently placed me at the bottom of the triage list. We considered going back home, as the blood had stopped and no more clots came up.

About two and a half hours later, after the drunken O-week student who had fallen over and bruised her shoulder had been sent off, I had a chest x-ray and was then ushered into the ED. They had finally called the renal registrar and it sounded like I was going to be admitted. I only spent the night, and managed to only cough up one more, old clot.

I am approaching 6 months post-diagnosis and I'm almost at the end of my course of cyclophosphamide (chemotherapy). My hair has thinned and is falling out and my skin is dry. I've had the last subcutaneous Zolodex implant, I'm weaning off the prenisolone and will move onto a maintenance drug in place of the chemo. My eGFR is currently at 30 and holding, my white blood cells are slowly dropping to normal levels, as is my Creatinine. My ANCA-PR3 antibodies are only just positive, I am not sure about the status of my GBM antibodies.

I'd like to think that I'm very lucky. One of the member of the renal team told us that if I'd left my symptoms another couple of days that I probably would've died.

I am now back at work, full-time, and have started playing netball again. I stick pretty closely to the renal dietary requirements (high protein, low sodium and low potassium) for my meals, which are quite different to Matt's dietary requirements (high salt, high protein) with his CF. I compare notes with my Dad about our kidney functions, medication and 'No fun' diets, as Dad's also a renal patient at Geelong. I now take medication to counter the effects of other medication, but I still don't take as many daily as Matt (13 vs 56, if you're counting. Half of Matt's are his digestive enzymes). The DVTs have resolved in both of my legs and i am now no longer injecting myself with blood thinners.

Matt still wins the competition for the most medication in our house, mine are in the little cup.

Late May, Matt and I had our first holiday together since this has all happened. Though I'm not as fit as I was before all of this happened, I was determined to climb to the top of Mt Oberon in Wilsons Promontory. It's graded as a hard walk because of how steep it is. It took almost 2hrs, but we eventually got there and enjoyed the views.

Five months post diagnosis – On top of Mt Oberon in Wilsons Promontory, capturing the views.