MY MPN Story

Before my treatment– always hungry, but still losing weight!

In the mid 2000's I began to notice low level symptoms, including feeling hot and having body aches and stiffness. I put the symptoms down to mild arthritis, ageing and a lack of physical fitness. I worked full time, considered myself healthy and rarely had time off sick. I started having a monthly remedial massage and began taking fish oil, glucosamine and Vitamin C. Over time, I forgot about how I had felt.

When we moved house in late in 2011, I thought it was about time I got on the books with a regular GP. In my previous suburb, it was almost impossible to get a regular GP so I'd see whoever was available or going to the part time casualty department at the small local hospital. As a health policy analyst I knew the importance of good primary care and passing 50, I was aware I was pushing my luck not having regular routine checks.

My new GP did a Full Blood Examination (FBE) as part of her base line assessment and found an elevated platelet level (in the 600's). She requested my history with the previous GP clinic, and to both of our surprise, there had been another FBE with high platelets at some point in the past. She immediately looked up references in her office and referred me for a blood test for JAK2. When it came back positive, I was referred to a private haematologist who diagnosed Essential Thombocythemia (ET).

Between 2012 and 2016, my platelets were steady in the 600's and I took 100mg of Aspirin daily. I was on 'watch and wait' seeing the haematologist once a year, having a blood test before I went. He would examine the size of my spleen, look at my results and send me on my way. Early on, he had mentioned that ET had a risk of transformation but I never really thought about this as anything other than a distant future possibility. At my 2017 check, my platelets returned to normal and I was taken off the Aspirin which further reinforced this view.

At my check up in September 2018, my red cell count was in the anaemic range at 105 and my spleen had become palpable. I had lost around 4 kgs in the previous couple of months despite maintaining a healthy appetite - which I was not that unhappy about! I generally felt and looked well. However, unlike my 15 minute appointments of previous years, there was a new tone to the discussion when he told me he suspected my marrow was becoming fibrotic, and I was reminded of previous conversations about 'progression'. I was handed the Leukaemia Foundation MPN booklet with the page on Myelofibrosis (MF) marked with an asterisk and I was almost frozen with shock.

I was advised that there was no cure for MF but with my anaemia, spleen enlargement and weight loss, I would be eligible for PBS funded Ruxolitnib to 'slow it' - subject to confirmation of MF in a bone marrow biopsy. I walked out with a referral for a gastroscopy and colonoscopy to eliminate 'other possible causes', and a new blood test for the anaemia, with another appointment set for 3 month's time .

The scopes were clear and I read everything I could about MF. I went back to my GP to talk about my concern at the haematologist's lack of urgency in doing the biopsy. I had previously worked on a Cancer services project in 2007 and a colleague from that time had been urging me to get a second opinion from the Haematology multidisciplinary team at the Peter MacCallum Cancer Centre (PMC). My GP tried persuaded me not to, re-assuring that 'doing things' was not always advisable and my haematologist had a history caring for me that would be missed in the public

system. Her final advice was "Stop Googling!" so I agreed to go back and talk to him about my worries at my scheduled December appointment.

At that appointment, I was told the follow up blood tests revealed an 'incidental finding' of a paraprotein 'IgM' that signalled different disease – a rare form of non-Hodgkin lymphoma (NHL) called Waldenstrom's Macroglobulinemia (WM). I clearly remember asking whether this was 'better or worse' than MF, and him hesitating briefly before saying 'better'. My immediate reaction was to be rather pleased – after all, Delta Goodrem had NHL and she got better! I asked again about a bone marrow biopsy, now thinking more about getting it in by the end of the year to avoid another $500 excess that I'd already stumped up for the scopes. Again advised that I 'was not sick enough' and that 'I should live my life as a well person and not as a sick person' and to come back in April 2019.

In the early weeks of 2019, my 90 year old father rapidly declined after pain in his thoracic spine was revealed to be a plasmacytoma myeloma – a form of blood cancer – and not the prostate cancer secondary originally thought. Over the same period, my weight dropped a further 5kg (and another clothing size) forcing me to buy new clothes. People I knew would continually comment on my weight, or just look aghast. Fatigue also became a real issue with leg cramps and night sweats waking me several times at night. I was also eating every 4-5 hours, including waking in the middle of the night hungry and having to eat.

I went back to my GP on Valentine's Day 2019, and insisted on a referral to the Haematology team at PMC, which she did on the spot. When I looked at my haematologist report to her, I was shocked to see that he had suspected 'co-existing' MF and WM – not 'either/or'. Within a fortnight of my referral to PMC, I had an appointment with an energetic young consultant who ordered CT and PET scans, a bone marrow biopsy and full genetic testing.

In the early hours of 1 April my father died and later that same day, I had the appointment at PMC to get the results of the testing. Both MPN and WM were confirmed and while both had similar symptoms, treatments were completely different. The next step was to try and work out which one was causing the symptoms and which one to treat first. The consultant suspected the lymphoproliferative disease was the most pressing current issue, as it was at Stage 4 and had infiltrated 80-90% of my bone marrow. He did assure me he was taking my case to the multidisciplinary team, at which Prof Constantine Tam (a world expert on WM) would provide advice.

In the following days it was recommended I have 6 cycles of combined monoclonal antibody and chemotherapy (Rituximab and Bendamustine) to treat the WM immediately and I was booked in for pre testing to ensure I could tolerate it. I started prophylactic antibiotics and antiviral medications and began my treatment on 1 May 2019. This involved 2 successive days of infusions at PMC, every 4 weeks. In my first treatment, I had a common side effect from the Rituximab that was like a severe infection, due to my antibodies being activated by the drug to attack the lymphoma cells. My Hb dropped as low as 89 before all my symptoms began to rapidly improve as my counts recovered. After 6 weeks I returned to work for 3 weeks in 4, taking my treatment week off. Towards the end I felt as good as I had in years (other than for minor nausea that was well controlled with medication).

I finished treatment in early October 2019 and the follow up bone marrow showed the WM in my marrow and blood was at negligible levels. The scarring had also greatly reduced allowing better visibility of the MPN. At this point my MPN diagnosis was changed from Post ET MF to PV.

Since then, I regained all the weight lost, but the low level symptoms I had years ago have re-emerged. My consultant described the two diseases as fighting each other to dominate with the MPN now holding sway. By January this year, my platelets had climbed to the 900's and my Hb to 180 – higher than they had ever been. I started Hydrea a month ago, with no noticeable impact on my symptoms so far. The platelets are responding (at 471 after 3 weeks) but the Hb is not, so venesection is a possibility.

Bottom line is that I am very glad to be at the PMC with its amazing expertise and resources and beautiful new building - and without the financial stress of gaps in cover for investigations and treatments in the private sector. The only thing I am really disappointed about is that I am not eligible for the numerous clinical trials available there, due to the presence of the two diseases!