

Paul's CMT Journey

Making the decision to amputate



Living with CMT

Hi, my name is Paul Thomas & I live in the Riverina in NSW. When I was a boy I had problems playing sport, running and so on. All my mates in summer used to have bare feet and I would be in shoes. One day we came across a pot hole in the road and they all said “hey Paul bet you can’t do this”, so they jumped in and out of the pot hole bare footed. I said there is no way I could do that. Secretly I wished I could, it would really play on me and yep, I am different,

At a young age I liked football so when it came to play for the town team I went to training for the first time and the coach at the time said “righto boys”, three laps of the oval. That was the start and end of my football career; I just could not run that far. I was a fairly active kid, I swam a lot and even swam in the town swimming team. I loved riding motor bikes and loved the river. I would sprain my ankle at least once a week but it was just normal for me it just didn’t hurt after a while. There was one time I was kicking the football and I put in for a huge kick and then I got a nasty pain in my right foot. I didn’t think much of it and just thought ‘oh well just another sprain’. After three weeks still thinking it was a sprain, I asked mum to take me to a doctor (I hated going to the Doctor) and yep it was broken. I was lucky that it had set back to the right place and eventually it was right.

How I found out and diagnosed with CMT

I was at college and I came home one weekend and my mother showed me this article in the Sydney Morning Herald, from memory the head line of the article was “Walking on Stones”. It read just like they were talking about me. Mum rang the number in the article and eventually was able to make an appointment made with Professor Garth Nicholson at Concord Hospital in Sydney. So away we go 8 ½ hour drive to the big smoke. We did test after test finally they hooked me up to the electricity test (I said to myself what is this guy doing) and eventually I

was diagnosed with CMT1A. Well least I know why I am a little different now. I think the doctor knew I had it when he did a couple of test as I know now I had classic symptoms. I cannot stand on my heels and I had no reflexes what so ever.

So life goes on and about 1999 I was living in Griffith and I joined up for a trial at Concord Hospital. Every three weeks I drove to Sydney for the research I was doing, night splinting and other exercises. After the trial my ankles were just sore and painful so I would strap them every day to go to work. I did this for a couple of years, I just wish we had known when I was a kid. I think that the night splinting would have worked. My right ankle turned out so far I got arthritis in the joint from rolling it so much as a kid. So I made my mind up I had to do something.

We did a bit of research especially through CMT Association and got in contact with Dr Grace Warren who had developed a procedure called Tendon Transfer. She recommended a doctor in Wagga Wagga. Off I go and see the doctor and as he was trained by Dr Warren this should be the sweet end of my pain we spoke about the operation on my ankle and also doing toe straightening. Boy was I wrong. The alarm bells should have went off when the doctor told me that he does not operate in the private system but I have another doctor that he was training and that he will be there for the operation. Big mistake!

The day of my operation I was very nervous. The new doctor came in to my room and said that everything is ok and ready to go but the original doctor can't make it. They gave me drugs to calm me down so I was not all there when I was wheeled into the operating room. The job done, the next thing I know I am back in my room about the same time I fully awoke. I was in that much pain, it was like a truck had run over my foot about 500 times. I pressed the morphine button 85 times in one hour. One of the nurses noticed how much I had pressed the button and said you must be in pain! The only relief I could get was to hang my foot out of bed and the nurses would go off at me for doing this, I think at one stage I must have cracked and said get the 'f ing' doctor in here and give me some drugs for a bloke my size not a five year old. The head nurse came in and they gave me a cocktail of drugs and I had no sleep at all. My head was spinning and still in so much pain.

The doctor finally came in the next morning. I had a back slab on my foot and it was so swollen and they took the back slab off. It took three nurses and the doctor to rip it off. I think that five staples from my wound came out at the same time. The pain was instantly gone. After I collected myself I asked the doctor how the op went and she said that we had to fuse the ankle.

I didn't think much of it until I spoke to mum, mind you I went in for a tendon transfer. Anyway I went home after a couple of days and everything was going well I could not believe that I had straight toes, I was that excited that I showed them off to all my family and friends. Have a look at the straightest toes in the south I only got the right side done. One of my friends gave birth so I went to the local hospital to say congratulations and it just happened to be raining and as it happens over I go on the crutches backwards. Natural instinct I put my foot down and yep it's the foot I had operated on three or four weeks earlier and by crickey it hurt. I rang the doctor straight away and she said is the back slab broken and I said no it all looks to be OK.

I went back to the doctor in Wagga Wagga, got an X-Ray and yep, the pins that they put in were screwing back out. I went back into surgery and they replaced the pins with bigger ones. I started all over again.

Three months passed and I thought 'if I had my ankle fused it should not move and the bloody thing is moving'! I booked into hospital again. This time I am in theatre for the x-ray so they can see what is happening and yep all the pins are moving around. There was talk of knocking me out and I got up and said there is no way that you are touching my ankle again. So I went back to my room and walked out of the hospital. I had lost all confidence in their so called expertise.

So what do I do now?

I got a referral to a doctor in Sydney. I went up and saw him and he was hesitating to work on me because of the work already done. I pleaded and pleaded and eventually he said yes.

Back to Sydney and I had another operation number three. Feeling great I stayed at my sisters for about six weeks, leg in plaster all the way up my thigh. That is the worst, not being able to bend your knee or scratch your leg. After getting the plaster off everything was looking good. I went and bought a new pair of shoes. Life is good at last and then everything started to move again. I immediately thought here goes round four, by this stage I had been on crutches for close to three years being off them for only a couple of months in that time.



My ankle just kept getting worse

Back in for another operation and everything went well and all on track.

I went to Melbourne and got a new pair of boots made for each foot - \$1300 later life is good and yep they were an ugly pair of boots but that did not worry me all I wanted was to walk.

By this stage my talis bone was gone and I had lost about five centimeters in height in my right leg. I had a built up shoe hence I had one pair of shoes.

The pins from this operation went through my from my heel, up my leg bone and believe it, this one came adrift. You could see it under my foot. I had to bore a hole, where the pin was pushing through, in my shoe and I wondered to myself how I could still walk.

Back to Sydney, had the pin removed and now five operations later the talk is about amputation. I said no and after a lot of discussion I asked if I could see another doctor, this time closer to home in Melbourne. He agreed and gave me the names of two Surgeons he recommended.

After making the appointments in Melbourne Mum & I went down, by this stage I was sick of doctors and their promises that they could fix my ankle. The first doctor wanted to put me in a

crow boot. I looked at mum and she knew that was not right. I don't know if you know what a crow boot is but it is a plastic leg strapped over your leg which totally encloses your leg. Where I live summer gets to 45 degrees, not an option. This could be great for some people just not me. The next doctor took one look and said "No Paul, I will not touch you but I have a colleague who you should see".

I finally had a chance. We made another appointment, back to Melbourne and what a great bloke he turned out to be. He never gave me a "yep I will fix you" but instead said we will have a try. I liked him and trusted him from day one. He explained everything so well, even having mum and myself look at a video showing exactly what was going to happen. As well as trying to fuse the ankle and he also was putting on a external fixation which surrounded my leg to below the knee all the way to my ankle. I had to have the bone cut just below the knee to allow for growth and every day I would turn the screw to extend the device a fraction at a time each day I would grow one millimeter. The idea was to try & get back some length in my leg which I had lost.

The recovery time for this one was ten months, non weight bearing & I ended up getting four centimeters back. I had nurses come to my home and do dressings every second day I had twenty four pins hanging out of my leg it took two and half hours to change the dressings we got that good with it we got it down to forty five minutes. The doctor did not want to push the limits so we were pretty happy with the results.

I finally got all the metal ware of my leg, the shin healed nicely as was my ankle. The next step was an AFO which I had fitted in Melbourne. It was great to be walking without crutches and for a while everything was going well.

Then my ankle started to collapse again.

By this stage in my life I had enough of operations. My Doctor in Melbourne said that we had two options, one, we can go in and try again this time taking a bone from somewhere in your body that has blood vessel and implant it in your ankle or two, amputation. I explained to the Doctor I had just had enough at the moment and I was really fairly well attached to my leg. I asked if I keep walking on my collapsed ankle would that jeopardize the next operation. He said it shouldn't as long as I could avoid getting an ulcer. So that was it. I will put up having a

buggered ankle and every few months I would get a new AFO made to account for the changing ankle.

Of course after a while an ulcer started, back to the Doctor again and after a while the ulcer improved. I eventually went to an orthotics place closer to home to get a new AFO and finally the ulcer cleared.

However, unfortunately other problems arose. It is now 2011 and I developed a severe allergic reaction on my foot The sole of my foot was just blisters, then my hands broke out, cracks started to appear and it got so bad I could not move my hands as the pain was a ten out of ten. This time I am back Melbourne seeing a Dermatologist Treatment included a very high dose of Prednisolone, applying wet bandaging every day and just about every steroid cream known to mankind,. It slowly started to improve so was referred to the Skin and Cancer Foundation for patch testing. After I had the patch tests on my back, I went off Prednisolone, from memory, for four days and before I knew it I had the biggest outbreak. Straight back on Prednisolone and as the results of the skin tests proved inconclusive but they believed I had a reaction to EVA, Luna soft material and also orthotic leather. This was exactly what my AFO was made from.. As I was unable to walk without the AFO I immediately had another one made this time out of carbon fiber. Over time the problem slowly subsided but never fully. It seemed like I was going to Melbourne every month for a couple of years seeing the Dermatologist but the best thing was I was still walking.

Then one day in late March 2013 I was driving to Melbourne yet again to see another Dermatologist I started to realise I had just had enough over the past couple of years and I really didn't want to continue down this path. So about 2 hours from Melbourne I rang my doctor and spoke to the receptionist and said "book me in for the amputation" We always had an understanding that both he & I would know when enough was enough. The first date was within three weeks which was too soon for me to get organized with work etc so the date was set for 22nd July 2013. I had made the decision, I was content with myself my next big hurdle was to telling my partner and family what I had just done. My Partner and family were very supportive with the decision, after all they knew only too well what I had been through and the pain I was in when walking.

Getting ready to lose a part of me.

Getting my thoughts together as to how to prepare for such dramatic surgery I spoke at length to my doctor and he expressed concerns as to my being mentally fit for this life changing operation. He talked over whether we should try and realign my left ankle, which was also deteriorating rapidly, first to help me have better support before amputating the right. Amputation won this argument, so he supported me all the way by putting me in touch with people who had had similar operations and always answering my questions honestly and with compassion. It was settled, I am to lose my right leg twenty centimetres below the knee.

Together we decided I would go to Bendigo for my rehab so after a phone call I had a meeting with the OT, Wound Management Team, Physio and the accommodation where I would be staying and, the soon to be the most important person, in the world the “leg man”.

So everything was a bit over whelming five or six people at once talking luckily my partner came to the meeting as well so she was talking to some and I was talking to the others.

I had a one on one with the Physio lady. I knew I had bad balance but not that bad. I failed just about every test that I did that day (in my head I kept thinking “what the hell am I doing”) & the Physio gave me a heap of exercises to do. I was determined to do them as best I could and I slowly I was getting a little better with the balance side of the exercises. All was looking good and the exercises they gave me were very low impact because of my weak left ankle.

The day before leaving for Melbourne for the operation.



Home modifications came next, luckily my partner’s dad is a builder and before we knew it, a new bathroom was built, hand rails, folding shower chair off the wall, rail in the toilet and new

steps to get into the house. It turned out to be quite a big job as one thing would turn into another thing. At the same time this was going on I decided I would have a 'legless party'. Two weeks before my operation I invited all my family and friends came along and we had a great evening. Some of my friends said that you should get your leg cut off more often and was even given a pair of "left socks" (I do have a wicked sense of humour and I always have a smile on my face)

Time has come

The morning I was going to Melbourne I saw my mum and to say good bye as I knew I wouldn't be back for a while. Up until this point I was strong, I had never cried, even with all the misfortunes that had happen to me over this journey, but that day the floods came. I was not nervous of the operation, just that I wouldn't be at home and work for a while. It was hard leaving.

My partner and a great friend set off for Melbourne where we stayed in a motel not far from the hospital, went out for tea & had a great night. All too soon it morning & day of the operation. I still wasn't nervous as I was mentally ready for this journey.

I booked into hospital and the nurse asked me to put the gown and compression stocking then I just waited. My doctor came in and we had a talk and he signed my leg"

this is the one". I said yep and jokingly I said can you please make sure that you cut it square. We all laughed.

Then the nurses came in and it was time to go. I said my good byes and away I went. A couple of hours later I woke up in recovery, very groggy. I looked down while the nurse was there and



I said I think we have a problem it was supposed to be left one the poor nurse went white, I started to laugh - she did see the funny side of it.

I woke up the next day and it felt great, no nagging pain in my ankle.

I was in hospital in Melbourne for eight days then a patient transfer vehicle turned up and away I went to Bendigo. I arrived in Bendigo at night and they put me in a room with another guy. I was not happy as I was supposed to have a room by myself. I stayed in that room for two days then I moved out to a Medi- hotel on the hospital grounds. I looked after my drugs each day. I was happy. Physio started and I was bandaging the stump myself. I was excellent at this! I was also helping another guy in rehab at the same time. I was going to the gym, and people who knew me were amazed as this was just not me but I loved it. I was that determined to get this over quickly and get home I put in 110%. It got to the stage that I was saying to the OT is that all you have, so each day I would try and break records that I did the day before. It seemed like such a long wait to get fitted for your first leg. It seemed like forever, especially when other amputees had got their legs and you could see the progress each day of what they were doing.

Finally my first new leg turned up I was like a kid in a candy store. The first time I put the leg on I thought that this going to hurt but nah it was right. I stood up & just to do that for the first time in twenty days was fantastic, no pain and I had forgotten how tall I actually was.

The OT's kept saying slow down but I was just so keen I asked the OT if I could take my leg back to where I was staying. I had to promise that I would not do too much which I did; I knew I had to stick to the rules. However sometimes you just have to extend the boundaries! My fantastic boss had bought me a new automatic ute for work, my partner drove it down to see me and away we went out to the bush. I jumped in and a way I went - I could drive , it was the best feeling.

Pretty much five weeks to the day I was walking unaided and back at work. I went to the RTA for my licence. I had to do a driving test and I passed. The next thing was to get in a truck so I got that licence back as well, now life is excellent.

Still to this day I am having trouble with the left leg, I'm still seeing a dermatologist and an immunologist & I hope that in the future I will overcome these reactions so I can get my left ankle looked at and try and preserve that side.

While writing this, it is my story and I'm thankful of all the wonderful people in my life so far and that I know everything happens for a reason. My decision would not suit everyone but for me it was my last resort.

I am blessed that I have never had a phantom pain, some things take a little longer to do, you have to be a little more careful but I think that I can do more now than I had been able to in the last ten years.

Now I have joined **Limbs 4 Life** as a peer support volunteer which I really enjoy when I am needed.

Thanks for reading my story.

Paul.



Living life to the full.