



Rachel – Cerebral palsy (and mum Margaret)

For Margaret Sutton and her 14 year old daughter Rachel, life is much more comfortable since Rachel was implanted with an intrathecal baclofen (ITB) pump. The anti-spasticity drug eases Rachel's tense muscle tones, making it much easier for the pair to manage every day activities, such as getting in and out of a wheelchair.

Rachel has normal intellect but little speech so mainly uses eye movement to communicate.

Margaret, a former children's nurse, adopted Rachel when she was two.

This is Margaret's story.

I adopted Rachel knowing that she had cerebral palsy.

I have two brothers in wheelchairs, one had a plane crash and one has spina bifida, so having people in my house in wheelchairs is no issue. I also have an identical twin sister who adopted an eight year old girl with cerebral palsy. We just thought that people in wheelchairs were the norm.

Rachel had been diagnosed with athetoid (writhing) cerebral palsy, characterised by involuntary unpredictable movements, very stiff at times and then floppy when tired or relaxed.

I attended a program at The Spastic Centre. When working with Rachel, I noticed that when she put any effort into her movements, these tight stiff tones would appear.

As Rachel got older I didn't know whether her tones actually got stronger, or whether she was getting bigger and I was finding it harder to manage her. It is a very difficult thing to really assess, her hands were flying everywhere. She had the typical athetoid movement, as everyone kept telling me.

Due to changes at The Spastic Centre where doctors were taken out of the system, I approached the Rehabilitation Unit at the Children's Hospital at Westmead to see what assistance they could give medically.

It was here where Rachel was diagnosed as having dystonia. When they explained what it was I suddenly understood that Rachel's cerebral palsy was not just athetoid but Rachel had a mixture of tones called dyskinesia. Her athetoid movement was one of Rachel's many movements, and dystonia was the major one.

When Rachel was upset, the stiff tones were dreadful and hard to manage and when she was tired or relaxed, she became very floppy.

Somebody explained that she was like a radio turned up too high or too low. There was no medium with Rachel. That made me understand a little bit more of Rachel's condition and her unpredictable movements.

I discovered that she could not access any home services, due to their 'no lift' policy, and using a hoist was impractical. I was left on my own.

Westmead Children's Hospital offered me a solution. They suggested that Rachel be involved in a clinical trial, where she would be given either an anti-spasticity drug or a placebo. After the trial Rachel was fortunate enough to be placed on oral drug, however the severity of Rachel's spasms meant that in order to relax them, the dose of the drug she required to relax her tones made her too drowsy to function in school.

Rachel's doctor told me about the pump. This required less anti-spasticity drug with a more direct effect.

The stiff tones hurt Rachel. They were so painful I could see it on her face as her body twisted. I don't think any mother knows what it is like until they've been there. The pump was one way out of a miserable and stressful situation.

Although the doctor explained that the implanted pump would be noticeable underneath the skin, Rachel and I both agreed that the cosmetic side of things was not as important as managing her stiff tones. Rachel agreed that beauty would not remain if her spine became twisted and her life was miserable.

There are always negatives, but they were minor when you considered what the positives might be. I knew that the hospital wouldn't have offered the therapy to Rachel unless they knew that she was a very good candidate.

Rachel was on oral drug for a year before the doctors offered the ITB trial.

Before the trial started it took two people to get Rachel in and out of her wheelchair. Within an hour or two of the trial, I could pick Rachel up and I could put her back into the wheelchair by myself. Rachel showed a very good response. She was pleased with the result.

We went in to have the pump inserted without any major issues. It was an operation that I think every parent would worry about – the pump goes in a pocket under the skin in the abdomen and a catheter runs from the pump into the spinal cavity. I had so much confidence in the Children's Hospital at Westmead and the people on the ward were just wonderful.

I think it took about six to nine months to adjust the right levels of the pump. We went up with the dosage and we came down and we've now come to a lovely level. The doctors had explained that it would take a while to adjust and every child is different, so I was quite happy with that.

I was delighted at how accessible her doctor made himself for the tweaking of Rachel's pump settings. She received an emergency procedure card that is tied to Rachel's wheelchair at all times, so I feel confident.

I don't have services sitting on my doorstep. I've found that having the pump, it's a lot easier to manage Rachel.

Rachel doesn't sit in her wheelchair all day, I lift her continually as she goes to the toilet four to five times a day, gets dressed, gets in and out of bed. All this puts a strain on my body. Now that Rachel is not extending as much, it puts less strain on me.

Rachel attends girl guides. She is a real socialite and our family is very much involved in community activities.

Rachel has stopped her jaw extension. She used to have very strong jaw extension and everyone would baby her because of it. Now it's not there. She's got a nice smile. She has photos without her mouth open.

She's become more verbal. She's able to answer, she can say yes when someone asks her questions. She's sleeping so much better through the night, so I also get more sleep.

Rachel looks more comfortable in bed. She just looks like a normal teenager.

Rachel is a happier girl with the pump and so much more relaxed.

Now that Rachel is a teenager I have asked her, "would it be better if we remove the pump?" Her immediate response is "no".