

Siobhan – our little miracle girl!

Richard & Carolyn have been married for 10 years and live in Brisbane; they have three children, Brianna, Matthew and Siobhan. Siobhan was born in 2004 and was diagnosed with Pituitary issues at around 4 months of age. This is her story.

During the pregnancy things progressed well. She obviously enjoyed her time in the womb for she certainly did not want to leave. On the 3rd May 2004 at 9:30am Carolyn's induction process was commenced with Syntocinon being eventually used to move things along and finally at 11:35pm on the 4th May Siobhan was born, term plus 14 days.

When she was eventually born she was not breathing. A quick trip from the labour ward down the corridor to the resuscitation room were several doctors and nurses suctioned her and cleaned her, each of them commenting on the Vernix, "she must be a premy", but at 42 weeks and weighing 3870gms (8lbs 5) she was definitely not.



Shortly after, Siobhan was returned to Carolyn in a Humicrib where she stayed for a short time whilst she was given additional oxygen. As with our other two children we noticed Siobhan starting to turn that rich orange colour that in our case was always associated with ABO blood incompatibility. As Siobhan's body begins to produce her own Type A blood it conflicts with Carolyn's Type O that was still present in her system. The dying cells result in Bilirubin being produced which the immature liver is not able to process. Siobhan became more orange and more lethargic; she was then placed into the special care section with 4 UV lights placed under and around her. The UV light treatment works by breaking down the Bilirubin, this process for Siobhan took 12 days and at times her levels were so high that blood transfusions were ordered, though not performed.



**Siobhan prior to light treatment
at 4 hours old**

At one point during this time her blood sugar levels dropped dangerously low and she stopped breathing, around this time with a failed IV and collapsed veins Siobhan was moved into intensive care, there Doctors performed surgery to insert a belly button IV. Siobhan stabilized and eventually we got to take her home after 14 days, though neither Carolyn nor Richard was convinced things were really right.

As in hospital, Siobhan did not show any interest in feeding, she could attach and suck fine but she would never stay there for long, it was as if she was satisfying a brief thirst but then didn't want to feed. Carolyn saw several lactation consultants, doctors and pediatricians and expressed her concerns. All noted she wasn't gaining weight but said she is well hydrated and put this failure to thrive down to being slow to recover from the blood incompatibility. Carolyn found it best to wake Siobhan just before she woke, no longer than 4 hours from her last feed and while still half sleeping she would just go into autopilot and seemed to feed a little better. Carolyn would also express off the fore milk before starting so she was getting richer milk, then expressing afterwards to keep up her supply.

Every so often Siobhan would vomit as she hit the full button; this was just devastating for Carolyn after working so hard and feeling like she had finally been able to get a decent amount of food into her.

At this stage Carolyn was being given all sorts of advice on what to do, one suggestion was to increase the period of time between feeds to allow her to become really hungry in the hope that she would feed better. On one occasion Siobhan was allowed to go for just over 5 hours without food, Carolyn couldn't rouse her to feed properly and she was limp, in hindsight she was probably in a hypoglycemic state. Carolyn never repeated this approach but rather continued to wake her at 4 hour intervals, with renewed conviction this was needed.

Siobhan's hospital pediatrician had noted that Siobhan's thyroid levels were lower than the normal range and that there was no response from the body to try and increase it, he felt this was probably her normal level. As she was approaching 3 months and still not doing well, only recently passing her birth weight, the Doctor wanted her to be put on formula and fed solids, thinking Carolyn's milk supply might be at fault. With all the additional expressing and food supplements such as Fenugreek and lots of homemade chicken soup Carolyn was a like a jersey cow, making lots of thick clotted creamy milk, she could have started a dairy, just Siobhan was not demanding much!

Carolyn had also tried bottle feeding her at home with expressed milk to see if she found that easier, but still Siobhan did not suck well from the bottle, she just rolled it around in her mouth, lots of different shaped and flow size teats were tried but it was quite useless.

Unhappy with her progress and with the one thing we knew that wasn't normal being the Thyroid, Carolyn sought a private referral to a Pediatric Endocrinologist. On seeing the paediatric endocrinologist he immediately said she had a Pituitary problem and put her on Thyroxine and after performing additional tests, Hydro-Cortisone.

Still failing to feed well, Carolyn tried a child health centre where they watched her feed and then tried to feed her additional breast milk after she had finished at the breast and they too could do no more than we were doing. They referred her also to a dietitian and recommended she start vitamin and iron supplements.

The results from an endoscopy showed no significant reasons for Siobhan's lack of feeding, just an immature valve at the top of the stomach which explained the vomiting when full. Siobhan was now given a Naso-gastric tube to increase her food intake. What a relief! As horrible as it was inserting the tube, finally we were able to be sure she was feeding well. Initially the dietitian got us to add a small amount of formula to the breast milk, but she gained weight so rapidly this was quickly removed. Our baby finally began to get chubby cheeks and some substance to her, but still no growth. From this point at 5 months until 18 months she became our little Michelin girl. She wore the same outfits for two seasons – talk about economical!

Due primarily to increased body weight from pump feeding and the fact she had tiny feet, Siobhan was close to 2 years of age before she started standing, she did however have a very quick bottom shuffle that amused many people, it was not however fast enough to escape the vacuum cleaner, for on one occasion she got too close and it caught her naso-gastric cord sucking it up and whipping it straight out of her nose – ouch!

Siobhan had a brain scan at 12 months which showed the entire anterior pituitary gland missing but a remnant of the posterior, which appears to be enough to control her electrolytes and water balance. It also showed she has elongated brain tonsils, but at present this does not affect her neurological function, though she should be watched if head injuries occurred or if she were to have sight or balance symptoms.

A Bone scan at 12 months showed her bone age as a "new born", no bone change from birth, but after the PBS finally approved the growth hormone at 18 months she started shooting up like a mushroom, in particular we remember the rapid change in her feet which till this point were the size of a new born but that seemed to double in size overnight. Earlier whilst in China on a work trip Richard tried to buy shoes for Siobhan. The shop assistant asked her age and Richard told her "she is just over one", she would not sell him the baby shoes, as they would in her eyes be too small, so he left empty handed.

These days she sits just over the 50 percentile on the growth charts and continues to grow and thrive like any other healthy little girl. When her bone scan was redone at 2 years it showed 6-9 months growth, then at age 3 it was 2 and again at 4 the bone age had increased to a 3 year old.

We have had a few scares with Siobhan at times when she gets sick and her blood sugar and cortisone levels drop. She has got to the point of altered consciousness and fixed dilated pupils and ended up rushed to hospital in an ambulance. Even things as simple as vomiting bugs, tonsillitis and chickenpox have ended up with a day or two in hospital. When our other children get sick our household goes into lockdown isolation mode. Siobhan has a low base cortisone level which when she is healthy is all she needs, but when she gets sick we now increase her dosage up to 6-8 times her normal (the standard treatment is usually 2-3 fold but this hasn't been enough in the past). Since this recent adjustment she has stayed well away from hospital whilst dealing with the usual minor health challenges. Her current Cortisone medication levels are 2mg + 1mg + 1mg daily and Thyroxine 50 micrograms x 5 days & 75 micrograms x 2 days a week and Growth Hormone of 0.5mg injected nightly.

Carolyn and Siobhan March 2009



It is hard to believe our little girl is now 5 at school and doing really well. She is a real little character, full of life and a delight to be with, though somewhat headstrong!

We would like to take the opportunity of thanking all the doctors, clinicians and nurses that have been so helpful to our little miracle girl, Thank you.

Copyright
Australian Pituitary Foundation Ltd
2009