WOLF HIRSCHHORN SYNDROME TRUST

Summer 2017 Newsletter

Registered Charity Number 1038219

Website address: www.whs4pminus.co.uk

Wolf Hirschhorn Syndrome Trust Weekend

It is now 3 months since our National Conference at Staverton Park and I have finally had time to write the newsletter. I can say that, for those who attended, it was a success in many ways. Staverton Park was a totally new venue for us having used the Hilton in Northampton in previous years. As with all new ideas there were good points and a few areas where we perhaps did not meet the expectations of some families.

You will appreciate all the hard work that was put into making this happen and big thanks goes to Sarah Fleming for all the hard work done over the past 2 years. Since the Conference we have already had a committee meeting, at which it was agreed, that we should start to organise the next conference in May 2019. Sarah has offered to organise this one for us but that will be her last. We therefore would like someone to step forward to shadow Sarah whilst she organises the next one.

It was decided to change the format this year and make the conference more fun but still informative. This I think we achieved with the 'Super Hero' theme. Well the compere certainly entered into the spirit of things! We had some speakers on Saturday. There was an informative talk on Epilepsy, part of which is shown later in the newsletter. However, the full presentation you will find on our website (www.whs4pminus.co.uk). Please do take a look. We had a talk on independent living, with a super hero theme, from Disability Rights and Dr Gerri Matthews-Smith updated us on the Hospital Passport that she is working on.

Dr Quarrell spoke to the newer families about WHS and genetics. Dr Quarrell is retiring soon after many years of research into WHS and other genetic disorders at Sheffield Children's Hospital and has supported us for a substantial number of years. We wish him luck in his retirement. The trustees and committee decided that we should invite Dr Quarrel to take on the honorary role of Genetic Consultant to the Wolf Hirschhorn Syndrome Trust. This he gladly accepted, so hopefully we will see him around at our Conferences for many years to come.

Dr Oliver Quarrell providing an update on research

Saturday afternoon I led a discussion around the future of the Trust. Prior to the weekend, a monkey survey was sent to all those attending the Conference. Approximately one third replied prior to the meeting but Margaret managed to ask some more families to participate. The results were then discussed by all those present. The overwhelming majority of responses were in favour of keeping the Trust going and to continue to provide support for the wider WHS community. It was at this point I raised the question of needing additional volunteers to help run the Trust. The present committee numbers have fallen over the last year or so for varying reasons. For those that did not attend and would like to participate in the survey please go to our website (www.whs4pminus.co.uk) and click on the Monkey Survey tab. It's quick and easy to do.

Late Saturday afternoon all the WHS young people entertained us with a splendid Parade to showcase all their wonderful Super Hero costumes. The evening was rounded off with an excellent Dinner followed by a Disco ably assisted by DJ Will.

On the Sunday morning we all enjoyed a Family Fun Day. Morris Dancers, varying children's activities, stalls and a grand raffle.

We are in the process of looking at how we run the Regions and what the role of the Regional Coordinator should be. We are also looking at the website and how we use it not only as a place of interest but also as a repository of information for both ourselves and for Hospitals. We understand the need to educate more of the medical profession about WHS and what it entails.

In the crèche preparing fancy dress outfits for the parade

The committee are looking for people who would like to volunteer their services/time to the Trust. We desperately need to find someone to produce a more regular newsletter. If that's you and you have the time, patience and creative flair then please let me know.

Our next Trustees meeting will be held on the 29th October so if you have any points to raise at this meeting then please let me know.

I hope you all enjoy the summer break, although perhaps not exactly the break some of you might be hoping for.

We have some highlights from the weekend to share with you.

Stephen

An article by Shona Wallace

National Meeting 2017

I knew when I booked our hotel room and bought the t-shirts that there was a possibility that we would not be able to attend the meeting but I still hoped to find a solution. Unfortunately Tommy couldn't take the time off and I didn't want to travel with Clare on my own. However if I am honest that is not the only reason. Although I have been attending meetings since 1993, and have made many friends, emotionally I could not have coped on my own.

The first meeting is difficult because you don't know what to expect and you look around at the children who all have different abilities and medical issues and you wonder what the future holds for your child. The next few meetings you look forward to meeting with friends and providing support to new families. Then you start to notice the families that are missing because their child is too ill to travel or because they have lost their child. Your perspective changes over the years.

Sharing because we need to be free to acknowledge that it is difficult and I know there are others who feel the same.

WHST Weekend Photographs

Pauline and Theo dancing

Prize giving

Sarah loving the animals including the snake!

Izzy having fun

WHST Weekend Photographs

Parachute and balloon fun for Adam and Josh

Harvey enjoying the dancing with the Morris Dancers on the Sunday morning

DJ Will

Ted and wonder girl Bex chilling out.

WHST Weekend Photographs

Tommy and his dad at the disco

WHST Fun Day

Photo with the Morris Dancers

Archie and Family dancing at the disco

National Meeting 2017 Evaluation

Monkey Survey – 17 responses - Overall Experience scored 9/10

The Venue – Staverton Park Hotel

The majority thought it was great – 12 out of 17 scored 8 and above out of 10

- Great space for all of the activities – couldn't have done that at the Hilton
- Bedrooms were good – family rooms were large and easily accessible to rest of the hotel
- Use of wet rooms as changing areas worked well although one family requested that the hoist be left in the bedroom and not taken back to reception to make it easier
- Bar area was good in the main hotel (more accessible than the Hilton)

Feedback from those who scored it low:

- *"Back to the Hilton. Staff is better trained. Much more confident. More drinks available once food ran out weren't topped up. No drinks at lunch time other than water. To spread out put on 1st floor rather than ground. To vast a building"*
- *"Return to old venue. This hotel was too large, long corridors, felt a little disconnected, plus the pool was too far away. Food not as good as Hilton."*

Speakers

- Epilepsy Research UK speaker very engaging and informative. Disability Rights UK speaker less so.
- Splitting Dr Quarrel's presentation out worked well so that not everyone heard the same presentation they've heard before.
- Although splitting the audience up made it feel empty in the main conference (especially as some had also gone bowling).

Children's activities

The majority thought it was great:

- *"Amazing things to do for the children. Thank you. Very enjoyable."*
- *"I think this was fabulous, a lot of thought went into this event. All was thought of and the siblings had lots of fun. Disco was fabulous and the fancy dress parade was great."*
- *"Very well thought out and planned. There was an activity for everyone, all very interesting,"*

Some didn't though:

- *"The superhero theme was only of interest to younger children and siblings of primary school age and below, and may have put off other families from attending the conference as there were definitely fewer people there this year."*
- *"Not suited to our sons interests"*

Family Fun Day

Majority loved the Family Fun Day too:

- *"A very well run and fun day out for us all. Will be coming next year if there is one."*
- *"Excellent, Full of new ideas and overall better than before. A big thanks to everyone. Enjoyed it very much."*
- *"Very good. Surprised at the turn out, glad I got to speak to more parents that day. It almost felt like a festival with so much going on and to do."*

A few that didn't

- *"I appreciate the effort that went into it, but personally I preferred the previous day"*
- *"Again only of interest to families of very young children, except the entertainment which was good for WHS children of all ages - Morris men, parachute games and Book bags"* Room for seating would be good so families can sit down to enjoy the entertainment.

Crèche

- The crèche was a nightmare 48 hours prior to the event so we were very lucky and fortunate to have a crèche at all. They did a really good job based on the short notice. I was impressed with how they ran it. I would recommend employing them earlier next time so that they have more time to plan and we are not left potentially without a crèche service.
- Overall though they did a great job.

Favourite part of the National Meeting

- *"Meeting other families"*
- *"The Disco, Spiderman, & the Fun Day with the Morris Dancers. We thought the food at the hotel was very good and the service was excellent."*
- *"Loved the Morris dancers and the live animals."*
- *"Meeting lots of new people. It was our first national meeting and won't be our last."*
- *"Learning an insight into other WHST family lives and letting the hair down."*
- *"We enjoyed Saturday. It particularly the evening meal and disco. We have been coming many years and the most important part for us is the time we get to catch up with old friends."*
- *"Talking to other parents and all the while having fun and relaxing while doing so."*

How would you improve it?

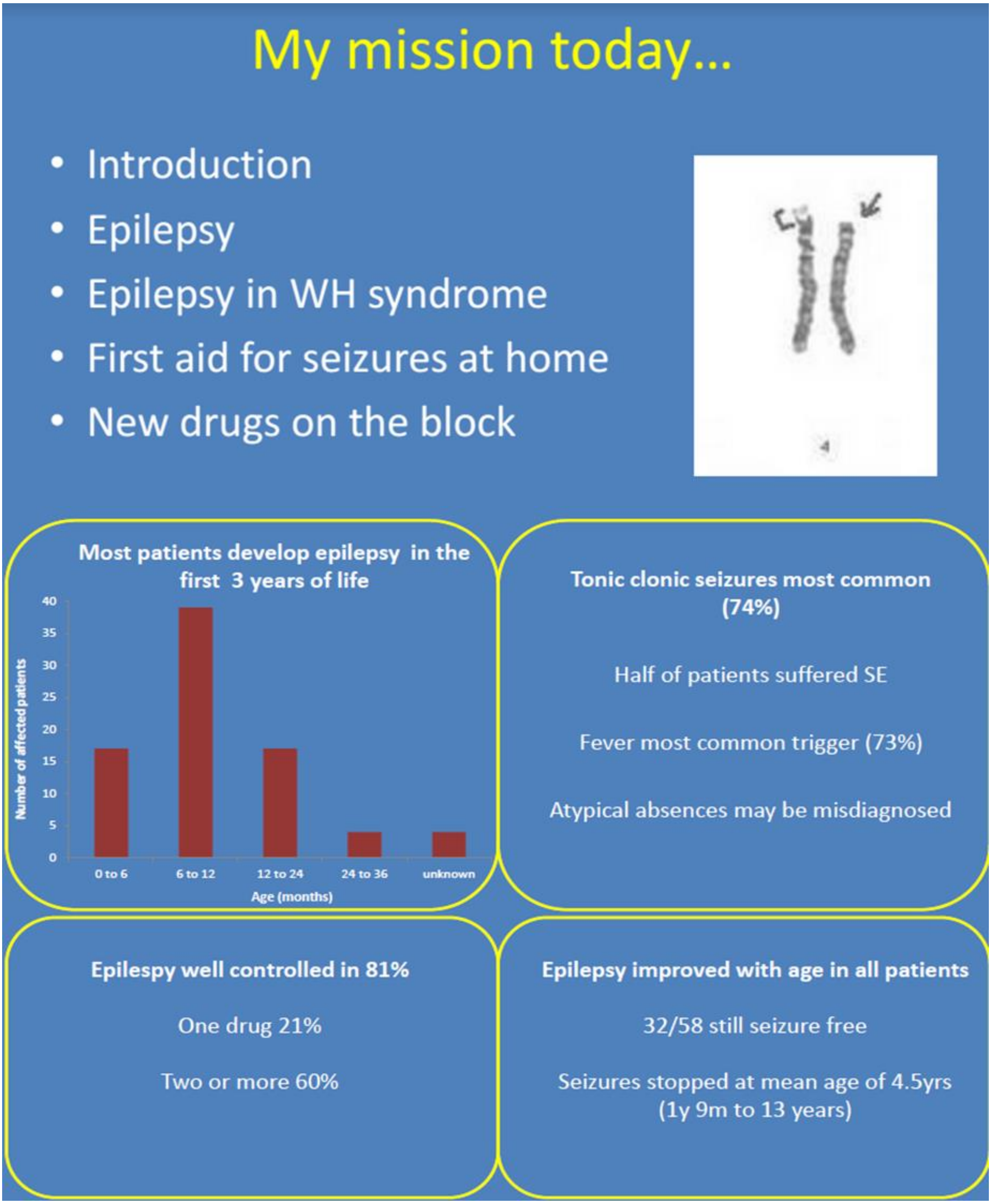
- *"A food / BBQ stall would be good. Maybe ice creams or perhaps join up with another rare diseases group to make the event even bigger."*
- *"It was really good. Nice to have a different venue this year, and we thought the idea of having a sort of village fete indoors was brilliant. Next time perhaps there could be more performers after the Morris Dancers...... live music, a clown, that type of thing"*
- *"More of the same please, it was really good. Why not include a children's entertainer to do tricks and sing songs etc. Also, how about fancy dress and a treasure hunt?"*
- *"Lectures on the Saturday could be mixed up a bit and a bit of trivia in between as sometimes there can be information overload. This is a weekend of learning, sharing and fun and overall a great event."*
- *"More activities for Carer's. Maybe pub quiz type thing. All WHS families given allocated area in restaurant. Instead of fun day, maybe day out to local attraction? The zoo?"*
- *"Maybe have more workshops, talks and demos given by i.e. therapists, other medical specialists"*
- *"Build on the new format! Good quality speakers and good entertainment via fun day for all."*

OTHER

- Schedule for the Saturday to be included in welcome pack or provided to attendees on the Friday, or emailed to attendees beforehand
- Theme worked well – recommend doing a different themed event for the next one
- T-shirts were popular
- Raffle prizes were great – more people to assist with sourcing prizes next time as a lot of the prizes were provided by our friends and families so we may not be able to obtain them again
- Photographer and compere were great. Compere added a lot to the energy of the event as well as a bit of humour. Feedback to me was great about both of them and a lot of people asked to pass on their thanks to them.

Speakers-Epilepsy UK

There was an interesting talk on Epilepsy given by Dr Sukhvir Wright of the Birmingham Children's Hospital. The slide show can be seen in full on the WHS website (www.whs4pminus.co.uk)



My mission today…

- Introduction
- Epilepsy
- Epilepsy in WH syndrome
- First aid for seizures at home
- New drugs on the block

Most patients develop epilepsy in the first 3 years of life

Tonic clonic seizures most common (74%)

Half of patients suffered SE

Fever most common trigger (73%)

Atypical absences may be misdiagnosed

Epilespy well controlled in 81%

One drug 21%

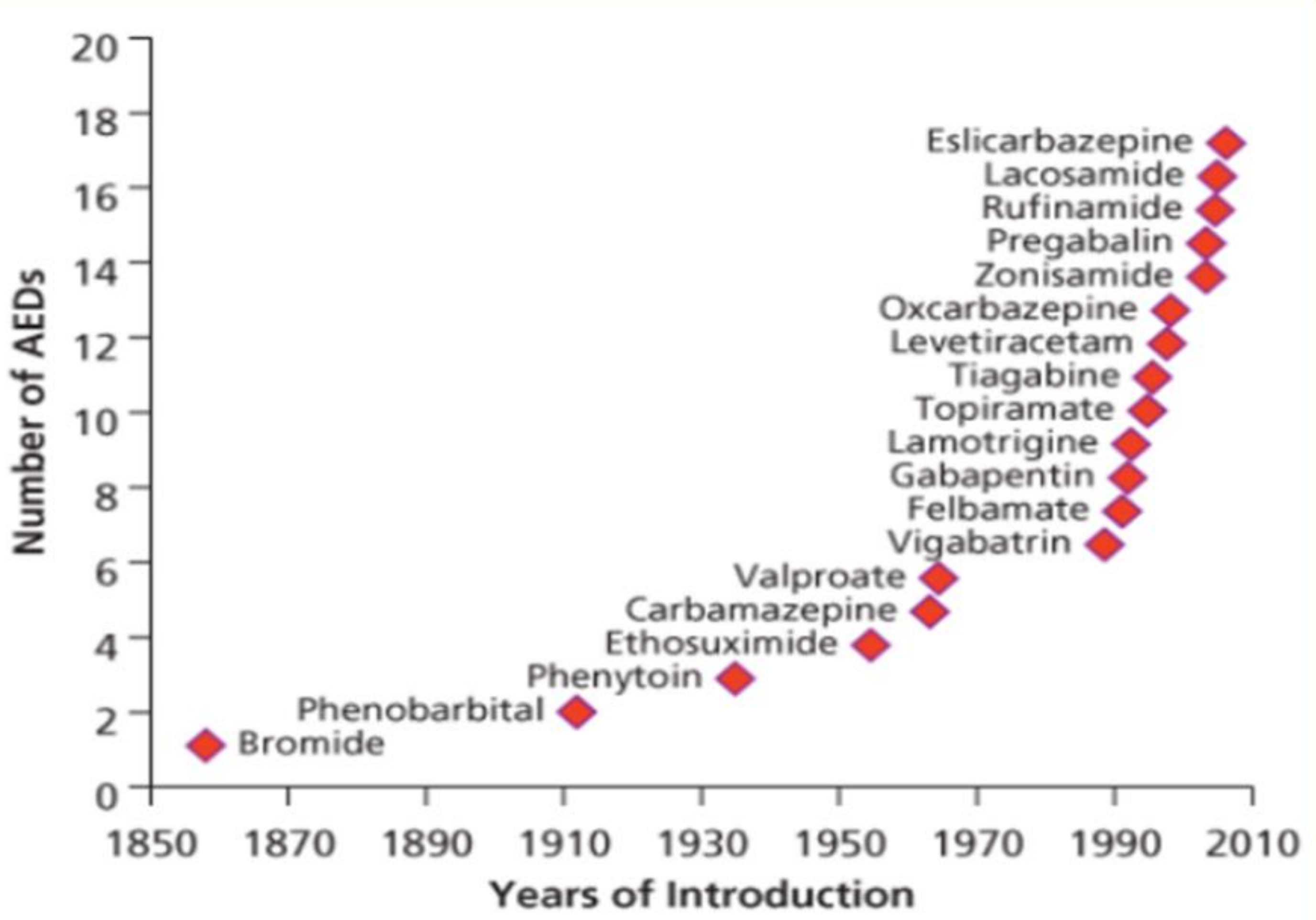
Two or more 60%

Epilepsy improved with age in all patients

32/58 still seizure free

Seizures stopped at mean age of 4.5yrs (1y 9m to 13 years)

Anti-epileptic medication



When the drugs don't work...

Ketogenic diet

- Mimics starvation in the diet
- Can be more effective that adding 2nd/3rd AED
- Requires commitment and support of a dedicated team (neurologist, dietician, epilepsy nurse)

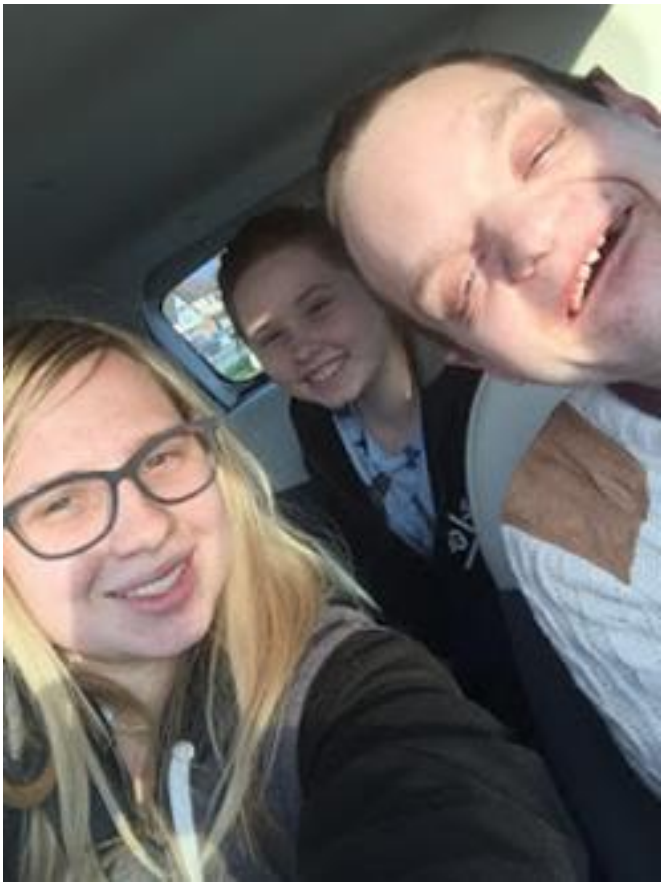
Article by Annelise, A sibling point of view

Tom's having Wolf Hirschhorn has rather obviously played a large part in both mine and Rach's life. I'm not going to discuss the profound impact it had on my parents - obviously their world was turned upside down when their first child was born and they didn't know what was wrong with him. They definitely didn't have the beginning of parenthood they were expecting.
I'm making this about the brothers and sisters - we are as much a victim to the condition as our sibling. And we always will be, no matter how old we get.

As myself and Rach are both younger than Tom we have never known any different - our brother communicates making childish noises, bum shuffles across the floor and continues to watch children's TV (I know most of the songs from the older Barney episodes and also know all of Dave Benson-Philips' Makaton video doing all of the signing along with it). My school friends were great and wouldn't say anything if they heard me sing "You Are Special" from a Barney episode I'd been subjected to at the weekend.

My parents did a brilliant job of not letting Tom's condition control our lives though. I had as much of a "normal" childhood as I was ever going to get. I was doing up to 24 hours of gymnastics a week, had various instrumental lessons, and went horse riding once a week. Rach went to dance lessons, also had flute lessons but didn't pick up the musical gene that I did, and also went horse riding... until she realised that she was allergic to horses and had to stop.

How did we manage to fit all of this in? As Tom got older, the more time he spent in supported living. At first he'd go for a day or two a week, and this continued to where we are now - he's in supported living full time at a house 20 minutes away with a group of boys who he'd grown up at both school and throughout his time in his respite houses. He comes home at the weekend and goes back on both Saturday and Sunday at 4pm.
In some respects, myself and Rach had to grow up quite quickly with Thomas. We had to learn very early on that life would not always be nice for our older brother. We had to learn to help him and our parents with things that none of our friends would ever get to experience. I mean what 14 year old wants to be able to say that they help give their brother his epilepsy medication and be on seizure watch, counting the length of each mini-seizure and the time between them? I remember doing this once and being in the car, writing down the times on my iPod touch while dad was driving us somewhere.



As I got older, I'd stay in at the weekends to help get Tom to bed - carrying him up the stairs got too much for my parents but I'd taught Tom how to get up the stairs himself. It's not something he'd do for anyone else, only me. It was quite often a challenge, but I love thinking about as I remember the laughs as I'd give him his milk halfway up the stairs and having "In The Navy" by the Muppets on repeat on his iPad. Quality brother/sister bonding time in an odd setting I will admit, but it was nice for what it was.

The siblings do have a hard time, there is no denying that. Do you know how often I have wished that my brother could just stand up and walk? It's the kind of thing that makes you have dreams at night, the kind of dreams that you just wish were real. Only the other night I dreamt that Tom stood up from his chair and rather wobbly walked to the bathroom to get something from the cupboard under the sink. I woke up and just stared at the ceiling wishing that a miracle would happen to take this all away from him. I want life to just give him a break.

Tom can't communicate with us so none of us have any idea if he is in discomfort, or he's ill, if he's just feeling a bit sad or just wants some time alone. We always have to play guessing games with him. All we have to go on are his cries or noises, which don't often tell us a lot. Mum tells that it isn't my job to worry about that – it's not my burden, but I do worry about it. And I always will. You can't expect me to not worry. I'd take it away from him in a heartbeat if I could, as I'm sure any parent would agree. I'd love to know that normal brothers are like but it's something I'll never have.

That being said, I'd like to think that the condition has helped me, as a sibling, to become the best kind of person I could ever be. I met with an old music teacher of mine a few weekends ago at our local music service's open day and she said to me, "It's so nice keeping up with you on Facebook posting things, in a world of so much hatred it is nice to see that there are still good people who care about those who would typically be left behind".

So thank you, Tom. As annoying as you can be at times you have taught me so much about life and I am eternally grateful.

An article by Annelise Begent

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