JOEL'S STORY

I would like to begin by thanking you for allowing me to share my experience with you all. You are all aware of the life-changing affects of reflex asystolic syncope (RAS) and in telling you my experience with my son Joel, I hope that it will give you assurance that others are going through what you are going through. I will tell you about my personal experience of having a child suffer from RAS from when it first occurred, how we got a positive diagnosis, how it has affected all our lives and the situation as it is today.

Joel was born on the 7/7/99 and is now 5 years old. He is the middle child having and older brother Aaron (7) and younger sister, Ellen, who is 2.

Joel was 5 months old when he had his first seizure. It occurred in a relatively normal situation. I had lifted him out of the bath and, on placing him on the bed, he went rigid for a few seconds, then went floppy. It was a huge shock to me, but I now realise that this was caused by the apparent shock of Joel coming out of a warm bath into the cooler air. Despite my original fears, I thought that as he looked and he recovered well, I would wait and see if there were any other signs of illness or any reoccurrence before raising alarms about Joel's health.

The next RAS occurred a few weeks after the first one-on Christmas Eve. I had left him with my parents and sister to go to the shop. When I arrived back home, it was immediately apparent that something was amiss when I was presented with three sets of tearful eyes and claims that Joel had awoke and when his granny had looked at him then he 'died' on them. This was very distressing for all, but again I thought there was no immediate panic. I would contact the health professionals. A few months later, in early March, a paediatrician reviewed him for a heart murmur and this was when the diagnosis of RAS was made. Joel was then 8 months old. I was assured that Joel would grow out of this at 18 months and he was discharged.

The RAS attacks continued. They occurred once every one week to three weeks, including when he had any invasive treatments such as vaccinations. When Joel was about 15 months I found a support group on the Internet. I joined the group. ***This has become my lifeline.***

At 18 months (the age he was supposed to stop having RAS) Joel had 10 days when he was having 2-3 attacks of RAS per day! During an episode Joel often received a secondary injury, such as falling against objects and sustaining minor injuries. However, unlike other sufferers, Joel has never slept after a RAS. Obviously, at this stage I became very concerned about Joel's health and development.

I shared my concerns with my GP and he referred Joel to another paediatrician. This consultant confirmed the diagnoses of RAS, and recommended that Joel be referred to a paediatric cardiologist for his opinion.

In March 2001. Again the diagnoses of RAS was given and confirmation of an innocent murmur. We were given a 24-hour monitor to record seizures but his episodes are very sporadic, and not surprisingly, he didn't have an RAS during a 24 hour monitor!!

Then we were given an event recorder and we were fascinated by the technology. We recorded 2 seizures and transferred the recordings via telephone to the cardiology laboratory. Joel's heart had stopped beating for 4 and 11 seconds. This was real progress, because now there was an explanation of why our son was blacking-out. We were reassured me that Joel would grow out of RAS by the time he would be 3 years of age. Joel continued to have 1 to 2 RAS weekly until August 1st and then there was a period from August until December were he had only 1 per month and he eventually had the longest gap of all - 6 weeks between RAS. We were very relieved that Joel's blackouts were reducing. He had just had his 2nd birthday, the situation was improving, and he seemed to be growing out of them. We longed for him to have his 3rd birthday. Luckily, throughout all this horrible time I was in contact with the support group via email, and getting information and support.

However, this happy situation was not to last. From January 02 Joel has experienced a difficult time. His RAS became more frequent, occurring once or twice a month. He has further associated problems, such as incontinent of urine during a RAS, and on two occasions we found him lying on the floor for no apparent reason. I sought reassurance from the patient group that this was connected to the RAS, since we were now worried about epilepsy.

At this time it became apparent that Joel required a tonsillectomy. He was booked in May to have the surgery and although the consultant knew about the RAS and supposed that this would not cause a problem for himself or the anaesthetist, however, on admission to hospital the anaesthetist expressed concerns about the frequency of the RAS and decided to speak with the cardiologist. The conclusions were that the hospital did not have the required cardiac medical team to deal with Joel if something untoward happened and the nursing staff felt they had little knowledge of the condition and were not confident about caring for him.

We accepted this as we certainly would not be putting Joel's life at risk and agreed to attend another hospital that had a specialist paediatric resuscitation team the following week. Joel had 2 doses of atropine prior to surgery, as one dose did not raise his heart rate sufficiently. His surgery was uneventful and he remained asleep for 2 hours and he had continuous heart monitoring, which the anaesthetist said showed he had a slow pulse rate.

Joel was never pain free from his surgery and 13 days after his surgery he was taken to the GP because there was some blood in his mouth. While in the surgery he had a massive haemorrhage and was rushed to the local hospital where he was almost unconscious on arrival. While in casualty I had to keep reminding the doctors of the RAS especially as he was for further surgery. The first anaesthetist had little knowledge of RAS and he got another anaesthetist to attend him. I was so worried that while the doctors where putting up the IV fluids and using many needles that Joel would suffer RAS during these procedures. However, thankfully he didn't and within 15 minutes of arriving at the hospital Joel was anaesthetised and having his 2nd operation in as many weeks. He lost a lot of blood and after surgery his haemoglobin was 7.2-half what it should have been. He was a lucky boy-he nearly died. He was discharged 4 days later, pale and weak but alive. He was only home 2 hours when he took a RAS. I really was scared. My main concern was the already low blood count and if this would result in oxygen starvation of the brain. I phoned the cardiologist and he asked me to bring Joel to see him the next day.

I explained the RAS were again more frequent about 2 – 3 per month and Joel was now being excluded from activities with his contemporaries. In addition, that Joel was also receiving a lot of secondary injuries during the episodes. It was affecting family life as family and friends were understandably reluctant to look after Joel. Another event recording showed Joel's heart had stopped beating for 12 seconds. The cardiologist recommended that Joel see a neurologist at the end of June 2nd.

The neurologist again confirmed RAS and although similar to the other consultants, is hoping he will grow out of it, he suggested trying Joel on atropine to prevent the RAS. Atropine is a drug that can block the effect of a nerve on the slowing of the heart.

Since starting atropine he has had long breaks from RAS of between 1 – 3 months. However, he has had numerous near misses, maybe between 1 – 3 per day. This has included dropping of the jaw and rolling of the eyes in conjunction with grey pallor and unsteadiness but followed by recovery so we know the atropine is preventing a full RAS. My greater concern has always been that Joel would suffer a secondary injury during an RAS. Joel's more recent RAS have led to secondary injuries. He recently sustained a head injury and bruising down the left side of his body when he had a seizure whilst standing upon steps and falling onto the concrete.

He was unable to use any protective mechanisms during the fall, which we all saw, as he was unconscious. The attack itself was triggered by being accidentally hit on the face with a football. Recently one weekend he had 2 seizures within 2 hours, both triggered by injuries sustained during play, both resulting in him falling again and banging his head. His 2 siblings witness these seizures and this impacts on them. The atropine has changed Joel's life. He is now able to play rough and tumble with the other kids without me scooping him up and taking him away from the kids. When he falls I don't worry as much that it will lead to a RAS. We have even taken him to the swimming pool, which he loves. Joel has regular medical reviews and is now in preschool, receiving extra supervision to maintain his safety, which is now being reviewed in preparation for him, entering primary school.

Joel is an active and adventurous 5 year old, who loves rugby and other sports and he is the age where he wants to join in such activities. We are now striking the balance between keeping him safe and allowing him space to grow up. Our hope is that his most recent attacks last week will be his last and that Joel will grow up enjoying the things that little boys enjoy.

As you can see RAS has impacted on Joel's health, social and educational development, as well as family life as a whole. He is 5 now and he continues to have RAS and near misses on a weekly basis. I want a better life for our son.

Since the patient group had been so supportive, it was only right that I should raise some money. So I sold the ballot tickets. The response to the ticket selling has been amazing! Everyone is so interested in the condition and has been so generous with their money. GP colleagues have mentioned people who had similar episodes which would probably have been RAS, in fact one GP now believes her younger brother may have had RAS as she remembers carrying her brother who was limp at the time outside to her mother, after either a shock or injury.

Joel has been a constant worry to us but the burden has been shared by the support given by others.

Cathrine Reid, Northern Ireland