Isabel

Isabel developed a rash on her knuckles in February 2008, just before her 2nd birthday. She also had red cheeks similar to slapped cheek syndrome, and a purple rash on her knees and elbows. At this time her temperament also deteriorated and she became generally grumpy, aggressive and miserable. A few weeks later at the beginning of March she stopped walking and preferred to be in her pushchair. She was also unable to get in the car, get up and down stairs, get into bed, or up and down off. If she did walk a few steps she had no balance and would wobble over flat on her the floor face as she could not put out her arms to save herself.

I was sent away from the GP with Vaseline and E45 for her knuckles initially, and was told when she stopped walking that she had a virus – after 2 weeks I demanded to be referred to hospital. We were sent to our local A & E which did blood tests revealing high levels of protein in her muscles, they consulted GOSH and we were referred there in March 2008 to the Rapid Assessment Neurological Unit (RANU). The consultant diagnosed her with Guillian Barre syndrome, a rare neurological disease which improves itself over time. Electrical reflex tests showed that messages were not getting through from her nerves to her muscles. By June 2008 there was no improvement and the consultant referred us to Rheumatology to rule out JDM. As soon as she was diagnosed with Guillian Barre syndrome we were referred locally for physiotherapy, which she attended twice a week.

When the possibility of JDM was broached, my local physiotherapist got in a colleague who was an expert in JDM cases, she took one look at Isabel and told me she had JDM and to expect a stay of several weeks in hospital. We attended her appointment at GOSH on July 17th 2008, and Isabel was admitted as an emergency. At this point she could not even roll over, and had a CMAS (muscle strength) of 9 out of 53. She was classed as a severe case of JDM. The doctors started her on IV steroids and Methotrexate immediately, and she commenced intensive physiotherapy at GOSH a few days later. We were released after about ten days and came in daily by ambulance for physiotherapy. After about a week we saw Dr Pilkington who said she was in danger of choking, and that the IV steroids had not worked. She was readmitted to the ward for 4 weeks as she started treatment with Cyclo, she had a total of 7 doses from August 2008 to January 2009. This, along with a total of 15 weeks intensive physiotherapy at GOSH, appeared to do the trick and she began to turn a corner in September. We were inpatients at GOSH from July to October, which was tough as we also have another daughter.

We think ourselves very lucky, as apart from some remaining muscle weakness and lack of stamina, she is now living a relatively normal life and attending nursery everyday. She has been off steroids since October 2009, and remains on a weekly injection of Methotrexate. We attend GOSH every 3 months as outpatients to monitor her progress. We do not know what the future holds, which is the biggest worry I as her mother have about JDM, but remain vigilant to symptoms and have blood tests every 4 weeks.