Wow – what a day!

The first meeting of "KLS Support UK" was held at the International Pediatric Sleep Association (IPSA 2012) Congress held in Manchester, December 2012 as a parent support afternoon prior to the main conference and was facilitated by Dr Cathie Hill, (our consultant) and had guest speakers Emmanuel Mignot and Isabelle Arnulf, a patient panel discussion and the opportunity to give Blood to Mali who had flown over from Stanford to fly blood samples back for research purposes. There was also the opportunity to meet up before the event for coffee and conversation, as well as join other KLS families for dinner in a nearby hotel.

This is my account of the day, some of the information I noted and would like to remember and my own interpretation of the things I heard. It may be accurate, but it may also be how I read and understand things from the people I listened and talked with. I believe KLS support UK will be sharing an official account of the day at some point in the future. http://kls-support.org.uk

We drove up to Manchester with the aim of meeting as many people with KLS as possible, we wanted to understand the condition more, hear about KLS and how it effects them, perhaps some things to avoid, and to meet some of the wonderful people that I had become friends with on Facebook. We knew that a new research paper had been published recently, we had already purchased a copy so anticipated that the medical professionals would be talking about this, and therefore did not expect an immediate answer.

Through Facebook we had spoken to other families that were going, my family had already met one other family with the condition, and I had already met 3 people on the board of KLS Support UK, so I had an advantage, I knew what some people looked like.

The first family we met in Costa made a big impression, our teenage son met another boy with KLS, the family had texted me when they arrived, and when we arrived at Costa they recognized me by my coat. Spotting people with an invisible condition is not easy, I then recognized someone from KLS Support UK and we joined them on a large table and introduced ourselves and others. Just over coffee I think we spoke to several different families who were affected by KLS including one family briefly whose daughter was asleep and in episode.

For me personally, I really appreciated this family in particular in coming, they had travelled the furthest. This was the first time I had seen a KLS episode in someone other than my son and although we did not chat long, I would say that this really made a lasting impression. Looking at this teenager, asleep cuddling into her mum and grandma, made me think, that is it, that is us! I have sat with a teenager asleep on me in a public place, tried to carry on as normal but in the back of my mind wondered if everyone was looking. When I scanned the cafe looking for other families I might recognize, I scanned faces, I did *not* notice a sleepy teenager cushioned in a parents lap, it was only later when I spotted a member of KLS Support UK talking to them that I noticed and then realized that they too had the condition. *"There was no imaginary beacon flashing over head saying look at us."* I have often wondered if that is what other people are doing…. Later in the day when we moved to the conference centre, this teenager had the same pale skin and vacant eyes that I recognize so well, I am disappointed I did not talk to them more, and disappointed for them that their daughter is unlikely to remember the day, but I would to meet the real teenager one day and see her personality as episodes just suck the patient's personality away.

Whilst having coffee, I met up with two members of KLS Support UK who Jake and I had met before, although Jake had no memory of that meeting, he was in episode and had struggled through the day. Last time they had meet a pale, tired, withdrawn individual, today they commented on how nice it was to meet Jake and how different he is, more confident,

bright eyed, vocal - I was pleased they had the opportunity to meet the real Jake that KLS takes away, and I definitely feel that having the opportunity to see another person in and out of episode helps us as parents and family, but also helps our children understand the condition.

I had been talking to another family prior to the conference who I was planning on meeting for coffee, they had an eventful journey, they left home that morning with their teenager in episode and she had woken up on the way. Her episode ended on route to Wednesday's conference, talking to her, this was the first time she had come out of episode other than at home, she was wearing clothes she did not even own the previous week, there was snow outside the car window, and when she woke she thought it was Friday. This amazing young lady just went with the flow, talked about her experience and some of the bizarre behaviours she had done in episode and managed to get her head round the experience so that she could talk with others. Her switch between episode and normality was just like Jake's and you wonder whether you should take him along in episode, but he can just switch at any point, if this teenager had been left behind this morning she would have missed meeting everyone, sharing her experience. You just never know when an episode will end in some cases of the condition.

Finally we headed across the road to the conference, unfortunately the morning session on another condition was running behind time, so we were allocated a room to mingle, we all had name badges on here, so I was determined to put some names to some faces, I met on the Internet and to share our combined knowledge of KLS....

The 1st Speaker – Emmanuel Mignot from Stanford Medical Centre – he had flown in from America an hour before he spoke. He shared some slides with us that we are not allowed to share but some of the key points I jotted down included:

- Exceptionally rare condition, 500 cases reported in the last 50 years.
- KLS tends to spontaneously disappear
- No known cause
- Some treatments can help a little, but no definite solution to KLS – no magic pill ☹
- The Future:
    - More Dr's need to be aware of the condition, people often miss diagnosed.
    - No one yet understands the evolution of the condition there is very little long term follow up.
- We looked at lots of tables and data and talked about probabilities – but there are very few samples to base the evidence on – Stanford have been researching KLS for about 7 years, have no current funding (will apply for some when they next discover something) and only about 500 blood samples to work with – with narcolepsy as a comparison they have been studying it for 25 years and have around 4,000 sample. The results included (amongst other things):
    - ¼ - ⅓ KLS patients had birth difficulties or developmental problems.
    - Possibly a genetic basis but no real patterns (data sample not large enough)
    - Possibly auto immune as in a virus attacked the brain but again nothing conclusive yet.
    - They are studying chromosomes and looking for abnormalities - but links could still be by chance. No definitive answer yet….
    - **To get results they need more blood from more patients.**
- Current research is looking into family relationships, testing for virus triggers (they are taking swab samples) and they are checking if autoimmune related. Current research is at the beginning of the journey – technology is improving, it takes time and it is hard with such a small sample. Current research is often done by students – once they find something, more funding can be applied for. Stanford will keep researching as you don't know

what the next results are due and whether those results are right – once they are proven and tested then they can be shared.

We then had the opportunity to mix, have a cup of tea and some of us gave blood. Then we went back in to meet the 2nd speaker. Isabelle Arnulf is based in Paris, France and who in 2004 worked with Mignot at Stanford and has since gone on to do her own research.

- Arnulf clarified the definition of KLS
  - At least 2 recurrent episodes of excessive sleepiness 2 days to several weeks in duration
  - Episodes are usually less that 18 months apart
  - Normal alertness, cognitive function, behavior and mood returns in-between
  - At least 1 of the follow – cognitive problems, derealisation, eating changes, disinhibited behavior is shown (could be kindness, rudeness, hypersexual etc)
  - Symptoms can not be explained by something else.
- With more awareness there are more cases, people are often misdiagnosed.
- Originally thought more boys than girls, since the tv program on Louisa Ball more girls have come forward. It often starts in teens, but not always, most have first episode in puberty, 1/3 had problems at birth, there are more episodes in winter and in most cases there is a trigger. Episodes peak when cold/flu is at its worse, no trigger connected with streptococcus.
- The sleep pattern of KLS patients is not normal. It is not the same as narcolepsy and the patient is not always asleep although they appear that way. Sleep patterns vary during the episode – sleep tests are not necessarily worth doing.
- There is cognitive impairment in 100% cases – apathy, 87% confused – with post-episode partial or complete memory loss. 94% slow to speak – we watch a video which showed patient gesturing to communicate – just like Jake.
- Differences were noted in handwriting, recording information, **Arnulf suggested there was no point in going to school or learning – cause they wont be able to read back what they have written correctly.**
- In episode they often can not – read, apply make up, cleanliness, shower, sports, social media, smoking, gaming with others…. There is also a sense of derealization in 100% of people – senses are less accurate, food tastes different, they don't feel temperature, they are unable to process information quickly – so to take a cross a road they can't hear and see the traffic at the same time, or to have a shower – they can see the water on their body, but don't feel it – have seen this with Jake.
- With time only the patient may know they are in episode – it is harder for the carers to tell,
- Other symptoms could include – wanting more sweets (for Jake it's ice cream), irritable, impolite, repetitive compulsory behaviors (this screams Jake is his OCD episode) and childish behavior (Jake every time).They can get depressed, anxious – 45% are scared of being left alone – they can get delusions, hallucinations, or be more affectionate – (Jake).
- Physically they can have a fever, are light sensitive (Jake prefers a darkened room) have headaches (yep – off the pain scale for Jake), sweat and feel sick.
- After short episodes there might be a brief overshoot with complete sleep loss for up to 3 days (we have done 36 hours awake), they are more chatty, feel relived and want to know what happened but they could also be ashamed, depressed and embarrassed.

- We look at pictures of brain scans and it was noted that most children with KLS were bright intelligent individuals. I noted that different areas of the brain were affected by different patients and in different parts of the episode.
- We then looked at how long KLS might last –
    - Her longest known episode is 16 months – I know people on Facebook who could challenge that…
    - There are less episodes over time, more frequent at the start but this can vary
    - The median duration is 10 years but can be longer if you develop KLS as a child or an adult.
    - 17% of patients sleep less well between episodes – they can be moody and eat more.
    - The patient's brain compensates for the episodes and rewires and they get good at doing this over time.
- How can we make life easier with KLS
    - Let them rest & sleep,
    - They will be rude and exhausted if prevented from sleep
    - They may fear contact with novelty, unknown people, may not want to be seen by friends and relations, they are also afraid of being left alone.
    - **The best things is to keep them home (rather than hospital), somewhere safe, a darken room under family supervision. Do not over stimulate the brain is suffering.**
    - Observe the patient – they could get suicidal but antidepressants will not help.
    - Don't let them drive – one patient has died.
    - **Hospitalize if major behavior outburst, severe delusions, high blood pressure, urine retention (but in episode they need less wee – 1 a day is fine!).** I immediate thought of one mum online who knowing this would have felt a lot better….
    - **When normal they may need a break after 30-45 mins. A pause during homework and exams could be helpful.** Will be sharing this one with school, he may get extra time in exams but they did not say this….
    - Do not put too much pressure on school and learning – if they cant they cant – I really think this one is easy to say than do when as a parent you want them to have the best future.
    - Relapses can occur after sleep deprivation, so have a regular sleep pattern, be aware of alcohol, sleep deprivation and colds combination or any of these…. Don't combined them if possible.
    - Drugs – no drug proven to work as KLS is unpredictable. When trying a drug it is a good idea to establish a baseline to see if things improve. If episodes are frequent it may be worth trying Lithium – I did not note much at this point it was hard to hear… but patients had to have regular blood tests and keep there levels at a particular level for it to work.

Well my brain is nearly at overload and we go to a parent patient panel, some of the discussion was relevant so I am only sharing what I recalled:

- Contact a Family is an organization who may be able to offer advice on education, the disability act can help get them to university.
- Lessons cant be done in episode
- School can help get a place at the next establishment by writing a reference/letter
- Lithium has to be taken at the same time every day – difficult to do in episode – many doctors's won't offer it. It is a toxic drug, it worked for some in the room and not others.
- Weekly blood tests in France can be done at weekends so they don't miss more of school if they are on lithium, so this may be available here.
- Episodes eventually ease off, there may just be overwhelming tiredness for a few days

- There can be days when you feel funny – the symptoms are less intense
- You have to learn not to overdo it as a patient, you may need lifestyle changes
- There are about 70 cases in the UK 2 cases per million.
- KLS UK first met in Feb 2011 – that is less than 2 years ago. The web site was launched in August 2011. The Louisa Ball program was repeated on television in August 2011 (and Jake had just started KLS and we never saw it – my brother in law saw a tiny clip and joked maybe Jake had it… if we had seen it we may have saved 4 months of not knowing). They are now a small charity and want to raise funds. They have attended some sleep conferences – which it costs them to attend and this has made more Dr's aware. They want to raise more awareness with professionals – there was a doctor in the room whose child had the condition and he had never heard of it – that needs to change. Raising awareness will help more families as some of the founding members of KLS had no diagnoses for a number of years

Without education Jake would not have been diagnosed when he did – Dr Cathie Hill, the facilitator had heard Arnulf speak before at a sleep conference – through her raised awareness she diagnosed Jake. Doctor's need to know – we only had a 4 month wait to get an answer.

We then moved back to the hotel – and were joined at our table for drinks by someone from Stanford. She had flown from America for a very short shop over literally to take blood samples back for research. She was able to talk about narcolepsy and KLS, she could not say which condition was worse, narcolepsy is continuous but currently has more treatment options, KLS you are more likely to grow out of, but because when it occurs you miss huge chunks of time, then go onto to live a normal life, those missing chunks could be during essential education, social and work years, it is hard to judge. She also talked about the importance of sharing stories, getting the media interested and creating awareness, the more people who are aware, the better, the more varied the stories are, the more inclusion of the range of symptoms the more likely they are to move away from the sleeping beauty stereo type. She also talked about the importance of making lifestyle changes - work hard, play hard and then more likely to be sick, a regular routine, planning ahead for late nights but having a rest before or a lay in afterwards or just a restful afternoon. Simple changes can help; do things sensibly and in moderation - simple non medicated solutions.

These are just some of the families we met during our time in Manchester, we believe there are around 50 cases in the UK at the moment:

A. Met in Costa, mum & teenage son with KLS - first boy with the condition my son met.
B. Teenage daughter in episode, mum and grandma, they had travelled the furthest and the hardest journey to get there and I am going to meet the non-episodic teenager one day
C. A couple from KLS support UK whose son did not want to come, but who was episode free since the summer, and whom spent a number of years battling the unknown.
D. A mum who only learnt about KLS this last week, so very new, and who child had a range of other medical problems too which were aggravated by the KLS.
E. A brief hello to daughter and dad, who were on the BBC program which I had missed when Jake was first diagnosed who had not had an episode for 18 months. I would have liked to talk to them more about fundraising for KLS but did not get the opportunity.
F. Mum, dad and daughter who had KLS as the result of a vaccination and who were the only named case in their country – I had read about them in the paper. Interestingly they were who were about to tell us that if it was law somewhere in Europe, then as part of the EU it had to be adopted here, so that is something I found really interesting, but might be worth investigating.

G. Mum, dad, teenage son and grandparents who had appeared on a regional tv programme here, the first male video clip I had seen. It was interesting to hear that their show had been shared in Europe.
H. A young lady and a boyfriend who I had spoken to on Facebook, a pretty confident person who like Jake had had loads of episodes since September and had only had the condition 2 months longer than Jake. I really admired her for coming and finding out what she could as I don't think I could had at that age, sadly she went into episode that evening, and was enable to share the day with her mum.
I. Mother and daughter, whose daughter was at college and had had KLS for a couple of months longer than Jake but not as many episodes – which highlighted every case is different, and whom I felt I did not spend enough time with.
J. A member of KLS support UK who's son has one long episode every year, who is trying to avoid one this year by removing the risk of infection. Inspiration to follow your instincts and try what you think is the right path for you.
K. Mum and daughter whose daughter has just completed university and got her degree, and whom had not had an episode in 20 months.
L. Mum, dad and daughter who we had met before and who we joined for breakfast. This is the third time our children had got together, this time they find it even easier to talk about the condition and share experiences. This family own a farm and can possibly offer a group of us to camp next year so the teenagers/adult can meet in a relaxed atmosphere, and if this happens I hope all my new friends can attend.
M. An older person who still has KLS, (it does not just effect teens and people in their twenties, just that age group in the majority of cases). His episodes are relatively short, he is aware of what is going on but has an overwhelming need to sleep. He is married, employed and has children, a great example that life can go on.
N. Son had KLS and mum works as a SENCO, she felt like I do, that there should be more information on KLS. I have agreed to talk to her about what our education plans are after the conference, and I think both of us would be delighted if at some point there could be some official guidance which education can refer to.
O. A mum and son who met someone with KLS for the first time the day prior to the conference. The son had not had an episode for 15 years, had a job, was newly married and had a young family. He had had episodes for five years and during that time he had 110 episodes. This gentleman shows us, is real proof that despite Jake having loads of episodes he can and will get through this. This amazing mum and son did it on their own... We have a whole team to support us .
P. Mum who traveled with her daughter and other family members who had all sorts of hurdles along the way today, her daughter left home in episode and who came out of episode on route who are still looking for a diagnose.

There were other families, but these 16 are ones I remember and who I would love to have the opportunity to meet again. These families may recognize themselves, some I had spoken to before, but all of them will make a huge impression as I feel each and everyone actually understands this condition and can offer us support and hope. I would still like the opportunity to meet others I have spoken to online, and to continue to meet these people again, because we have returned home with a real sense if belonging, there is a community that understand and a wealth of support available from each other as long as we are all brave enough to talk about it.

I would love to thank Dr Cathie for taking us on, for her involvement in the team that enabled this conference to happen for parents, and to KLS Support UK, particularly families L and K for making it happen.

And finally a reminder the views I have expressed are mine – they may not be accurate, but they are my memories of a day which made a big impression.