The second meeting of "KLS Support UK"

The second meeting of "KLS Support UK" was held the Governors' Hall, St Thomas' Hospital, London in September 2013. The guest speakers included Dr Michael Farquhar (Consultant in Children's Sleep Medicine) and Dr Guy Lechsziner (Consultant Neurologist) of The Lifespan Sleep Medicine Group at Guy's and St Thomas' Hospital. There was also short talk on educational issues around KLS by Sarah McKimm of IPSEA. There was also the opportunity to meet up during the event for coffee and conversation, as well as join other KLS families for coffee at a nearby Costa.

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 went to London with the aim of meeting as many people with KLS as possible, reacquainting ourselves with families we met last year, we wanted to understand the condition more, hear about KLS and how it effects other people , perhaps some things to avoid. We were interested in what the speakers from St Thomas's hospital had to say, wanted to see if they share any different information and as we are currently fighting our own battle with education we hoped that IPSEA could offer as some advice.

Last time through Facebook we had arranged to meet up before hand at Costa and stay in the same hotel with people travelling from afar, this time KLS Support UK had requested that I did not run a Facebook event for this and there weren't any discussion online about this opportunity – although three families did manage to meet us for coffee. It was lovely that Jake was able to chat with a KLS patient who had just completed year 11, and another in year 10 in a relaxed atmosphere so that he choose to sit with them in the conference. It was also great to meet with another patient who although had other medical issues going on understood KLS, was still having episodes but who was just starting his second degree. A positive and encouraging role model for the teenagers helping them realize a bright future is possible.

Dr Mike Farquhar

Our first speaker was Dr Mike Farquhar who shared the following information with us:

The text book description on KLS is:

- Hypersomnia averaging 2-31 days, with about 18 hours sleep,
- Hyperphagia
- Hypersexuality

Dr Mike was aware of around 500 cases of KLS in the last 50 years that had been documented with more patients in France – 1.5 per million there, he thought there was around 100 people in the UK with KLS

and around 25 of those they were aware of through the clinic. KLS is more common in boys and the median onset in around 15.7. He felt that there was an over emphases on the hyperphagia and hyper sexuality.

Appetite is an issue, 66% of KLS eat more, 34% have deceased appétit and 37% eat automatically. 53% show sexual disinhibition, more in boys than girls.

In some cases of KLS it is actually menstruation related hypersomnia and this needs to be ruled out.

Dr Mike felt there should be more emphasis on the Cognitive changes during a KLS episode – as patient may have derealisation, talk like a two year old, struggle to read, feel they are in a bubble or have increased slowness – an overshoot of KLS episodes is depression – lowering of the mood when patients are less tired then a high or period of agitation or insomnia before they return back to normal.

Dr Mike explained that the vast majority seemed OK after KLS but they really need more clarification on this and a larger sample of patients.

Dr Mike explained that they were working on clarifying a criteria for be able to diagnose clearly a case of KLS to be known as the ICSD-3 Criteria (2013) with the following 5 key points.

A. At least 2 recurrent episodes of excessive sleepiness of 2 days to several weeks
B. Episodes recur at least 1 per 18 months
C. Normal alertness, cognitivie function, behavior and mood between episodes
D. At least one of these during an episode:
    - cognitive dysfunction
    - altered perception, derealisation
    - eating disorder (anorexia or hyperphagia)
    - disinhibited behavior (such as hypersexuality)
E. Symptoms not better explained by other disorders.

Dr Mike then talked about how long the disease would last – something I would love to know but for us the figures he shared were not encouraging…

- Onset can be hours or days
- Average number of episodes per patient is 19
- The mean duration of episodes is 5.7 months
- The median duration of symptoms is 13.6 years
- Burns out?

He then shared with us a slide about treatment - he currently felt there was no effective treatment for KLS, it is hard to tell if the drugs work or it a gap/end of episodes is naturally occurring, he did suggest that the experts were the families with KLS, as we see the whole package whereas as a Doctor you only get a glimpse and that they did not often see a patient at their worst. **He suggested if you were considering trying a treatment – if you had less than 4 episodes in a year it would be difficult to gauge**

if it worked, if you had more than 4 episodes then it was worth trying something. He felt that if you then had more than 2 episodes in 6 months then the treatment most likely was not working.

He then went on to explain a pragmatic treatment approach, all of which I was aware of:

- Good sleep hygiene,
- Avoid Alcohol
- Vaccinate, treat infections
- Avoid weight gain
- Stay at home during an episode
- Don't try to stimulate
- Don't drive – hide the keys if needed
- Reassure the person
- Occasional symptomatic need to medication.

He then talked about what was happening with current research and stated that they have found no single gene as a trigger for KLS, and that they were looking forward to agreeing a case definition for KLS which would make the research easier, they wanted to look at whether there was a consensus in brain changes, whether these continued after episodes stopped, what the treatment options could be, whether there were any triggers, whether a generic cause was probable and then hopefully one day a cure – this all is a long way off.

Dr Mike then said that on the whole KLS episodes decrease with age, but in some cases they may last 35 years or more – when pressed whether teenage onset KLS would be outgrown by the time the young person is 30 he could not say it would stop.

Dr Guy Lechsziner

We were then introduced to Dr Guy Lechsziner – he gave us some more information about KLS. He admitted that they were no experts in KLS, most "experts" saw 2 or 3 cases of KLS in their lifetime and as they did not see enough patients it was difficult to get a full picture of the condition. When it came to diagnosing KLS, there are no known treatments that work so doctors do not want to make a diagnoses of KLS as if they can diagnoses something else that they can treat they would rather do that. A diagnoses of KLS is academic – they have to rule out everything else first, if they could say 100% for sure someone had KLS then they would have more text book cases to compare. He also explained that the delay in diagnoses was often due to the episodic nature of the condition.

He then told us about the long term aim at St Thomas's to set up a clinic with adult and child services for KLS so they can expand and educate doctors – they want it to take on a multi agency approach and will start with the patients already being seen there and then a central hub for other doctors working with KLS patients, which sounded very exciting. They wanted it to be a "one stop shop" to assess and manage KLS, to work with Prof Mignot and to have the aim be able to follow up patients in the long term too.

Dr Guy also gave some examples of why their first step was to get an accurate diagnoses criteria for the condition because without that they would not know whether they were treating KLS or something else, he citied an example here of a women suffering KLS like symptoms he was able to stop the episodes continuing with a migraine pill – but he could not say in this case whether the patient had KLS or not.

Sarah McKimm of IPSEA - www.ipsea.org.uk

Sarah spoke to the conference about how KLS provided unusual problems in education and it would be a good idea to try and get a statement from your Local Education Authority as KLS has some really unique issues and education needs to adjusted in the way it is delivered through additional services.

The education plans in the UK system are changing so that they apply to young people from the birth to 25 years – this might mean a watering down of services, but it means that services will need to be provided to support young people with KLS for longer which is important with the fact that KLS oftens comes on in teens and is often undiagnosed for the first couple of years – but importantly you do not need a diagnoses to start accessing the statement process.

When we pressed KLS Support UK and IPSEA if they knew of a KLS family who had got a statement – they did not know of one and IPSEA admitted that it was only recently she had hear of the condition. We are mid way through this process and know now Jake is going to be statemented – so possibly the first KLS patient in the UK to get this support so we would be happy to share this information with anyone who is going to go down the same route.

The KLS panel, which consisted of Dr Michael Farquhar , ,Dr Guy Lechsziner, , Sarah McKimm our speakers, Lily and Poppy who have KLS, Annette, Adele, Beverley and Caron the team at KLS Support UK and some of the discussions included:

- Record keeping – keeping notes of when episodes are on a chart so you can see if they repeat in waves and also whether there is a pattern or if things change over time.
- Living away from home – at Uni, with friends etc – it is important to tell people that you have KLS even if you don't want to, cause that way if you act out of character or suddenly start sleeping a lot they can contact your parents who know how to care for you, you can be collected taken home and they can put things on hold for you so you don't miss out.
- A special consideration for University may be extra time – one patient gave an example of having a year free cause he missed his exams due to KLS whereas another patient had to pay for those.
- The KLS'ers who had not had an episode for a while agreed that the last episodes were often weaker, sometimes just days of extreme tiredness and then it just went away.

Jake was presented at the conference with a certificate for the money he has raised with the support of our friends for KLS Support UK and this can now be seen on our web site.

KLS Families

In Manchester we met up with families A to P – it was great to meet some of them again... (I have referred to last years account so it kind of update….
http://www.kleinelevinsyndrome.co.uk/Documents/KLS%20Conference%20Manchester%20Notes.pdf)

A. .
B. .
C. A couple from KLS support UK whom spent a number of years battling the unknown. Son is now at University and doing OK – still has episodes but not for a while…
D. .
E. .
F. Mum, dad and daughter who had KLS, daughter had now completed school and was doing well despite KLS and who since had met other families who also felt vaccines were a trigger.
G. .
H. .
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. She was doing well had had a slightly longer episode and had to retake some exams cause she missed them due to KLS.
J. A member of KLS support UK who's son has one long episode every year, who was able to report he had not had an episode in 12 months, and who were looking at Uni choices for the next academic year.
K. .

L. Mum and daughter who we had met before five times, who had not had a full episode since November and was looking at going to sixth form.
M. An older person who still has KLS, who we have met since and who is sharing our leaflets to increase awareness of the condition.
N. .
O. .
P. Mum and daughter whom we met again, having episodes similar in frequency to Jake and whom has finally heard she might have a diagnoses.

New Families

Q. A young man and his girl friend with KLS and other complex medical conditions doing a degree and who share that he got an extra year at college free because of medical circumstances and reasonable adjustments. They joined us for coffee.
R. Mother and daughter from France who are keen to share information across countries.
S. A young lady with KLS and her mum who were in a recent newspaper article about KLS and graduating college, the article did not say all they wanted it to say but it did help with awareness.
T. A young adult who developed the condition at 18 who had trained to be a nurse and who gave the advice of the importance of telling your friends around you at college that you have the condition – cause when you are in episode you just want to be at home.
U. A KLS patient in her 40's who had KLS before she was 30 and whom was happy to say she was KLS free.
V. A KLS parent, a new board member and her 18 year old son who specialized in rare disorders and who really wanted answers to what causes KLS.

There were about 17 families there this time – a lot less than last time but the experience was just as valued and important, it is great to meet other families, share information, hear what they have achieved and to learn from them. I wish more families who have outgrown KLS would stay involved and share their experiences as the young people really need to know there is life beyond KLS.

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