KLS Support UK
Helping families affected by Kleine-Levin Syndrome
www.kls-support.org.uk

Newsletter 11 December 2018

We wish you Season's Greetings and a Happy New Year. We hope you and your family are well and episode free and for those in an episode we hope that the episode will end very soon. We would like to take this opportunity to thank all those who have supported KLS Support UK this year through fundraising, donating and raising awareness of KLS.

Save the Date 8 September 2019

The KLS Support UK Family Day will be held in London on 8 September 2019.

Report on 7th KLS Support UK Family Day 2018

Thank you to all those KLS families who attended this year's Family Day. Around 40 people came from all over the UK. We would very much like to thank our speakers, Dr Guy Leschziner, Consultant Neurologist and Lead Sleep Specialist at Guy's Hospital and one of our medical advisers and Dr Laura Madeley, clinical psychologist in the sleep disorder service at Guy's Hospital.

Dr Leschziner has this year written a book on sleep disorders, The Nocturnal Brain, which will be out in March and includes a chapter on KLS.

Dr Leschziner was invited to speak at the KLS Foundation International conference in June and took part in both the general sessions on KLS and treatments and the researchers' sessions where researchers and specialists from Stanford, Atlanta, France, Sweden, Taiwan and China met to discuss the latest research and diagnostic and treatment information on KLS. Slides of the talks at the KLS Foundation Conference.

Dr Mignot, Dr Leschziner and Dr Landtblom at the KLS Foundation Conference

Dr Guy Leschziner's talk

Slides of Dr Leschziner's talk can be found on the KLS Support UK website.

Dr Leschziner welcomed everyone although he said he was upset to see some of the same people as he always hopes they will have grown out of KLS since the last family day.

Dr Leschziner spoke about the KLS Foundation Conference meeting of researchers and said that there was now a glimmer of hope as to understanding what causes KLS.

He talked about Dr Isabel Arnulf's Paris KLS service, the largest in the world with 220 patients diagnosed with KLS and 20 new patients a year. The following advice comes from Dr Arnulf's clinical experience and 2 clinical trials.

Advice for during a bout

Stay at home

Limit noise

Remove phones

Don’t try to keep the patient awake

Do not drive

Liaise with school and work and make sure they know about the diagnosis and make allowances.

Do not use stimulant drugs – they do not work. Modafinil may keep a patient awake but will not help with cognition

Manage headaches with standard painkillers or Diamox

Once diagnosed, only go to hospital in the rare cases where treatment is necessary for management of autonomic dysfunction or if antipsychotic medication is required.

Long Bouts

1in 4 people have bouts of over 30 days. For those with long bouts the Paris clinic try the following medications.

Amantadine. Dr Leschziner is not yet convinced this does much.

IV Steroids - Methylprednisolone – Dr Arnulf found that when given, the episodes were shorter than previous episodes. If given in the first 10 days of an episode then they were most likely to work. If given within 6 days there was a 50% chance of shortening the episode by 11 days. Potential side effects include insomnia, muscle pain, tremors, headaches and possible major hip problem. Dr Leschziner believes the use of IV Steroids needs careful consideration.

In between bouts

Around 15% of patients have cognitive impairment, reduced attention, reduced verbal fluency and sleep more than normal between bouts.

Preventing relapses

Avoid infection - get the flu vaccination

Avoid alcohol

Have a regular sleep pattern

Prophylaxis - these try and prevent the frequency and intensity of episodes. Dr Leschziner said there are anecdotal reports of drastic response but no conclusive evidence yet.

Sodium Valproate – this is not allowed in the UK for women.

Lithium - Paris case study - 71 patients (only those with severe episodes) took lithium. It appeared to reduce frequency from an average of 4 episodes per year to 2 episodes, however the reduction was also seen in those not taking the lithium. The lithium also appeared to reduce the length of the episodes from 18 to 10 and 40 to 20 days. The Paris study found 37% reported having no episodes when taking lithium, 51% had reduction in episodes and 12% had no response.

40% of patients who take lithium experience tremors. It also can cause people to drink more and urinate more. Taking lithium needs careful monitoring.

Dr Leschziner said that other countries had not found lithium to be as effective as Paris presented. If a patient has very bad KLS then lithium may be worth trying.

Understanding the origins of KLS

Dr Leschziner showed a map of the worldwide distribution of KLS. There is a cluster in Israel with 23 people having KLS in a population of 7,000,000. It implies genes are at play,

4 families in the world are known to have more than one member with KLS. 2 families have siblings with KLS, one has a father and son and one has cousins.

Dr Leschziner asked if we knew of any family in the UK with more than one member having KLS. We are not aware of a family with more than one member with a confirmed diagnosis of KLS. If you are aware of families with more than one member with KLS please let us know.

Dr Emmanuel Mignot of Stanford University, who unravelled the origin of narcolepsy, is also heavily involved in researching KLS. He is working on a genetic study looking at variants in the genome. He is also looking at significant differences between those with and without KLS.

One variant identified by Dr Mignot in KLS is TRANK 1 which relates to how the nervous system develops and this variant is also seen in those suffering with bipolar disorder.

Another important finding is that 25% of KLS patients were likely to have suffered difficult births.

Dr Leschziner said that if you have donated your blood to the Guy's and St Thomas' Sleep Clinic your sample will have been sent to Stanford to be included in this research.

Dr Ambati of Stanford has compared spinal fluid in and out of episode. It appears the range of proteins changed in episode. Dr Leschziner explained that CFS proteins change when people are unwell and asleep so it is difficult at this stage to argue the causative role in causing KLS.

The change in proteins appears to show an immune response when in episode which would tie in with Paris' thinking that there is an autoimmune response.

Dr David Rye of Emory University believes there may be some shared features between KLS, hypersomnia and narcolepsy without cataplexy. Recognising the shared features are important as it may open up different treatments. Dr Rye prescribes Clarithromycin for KLS but Dr Leschziner is concerned about side effects including insomnia. It is possibly an option and may work for a select few, as there are reports it has worked for some KLS patients.

Future Directions of Research

Dr Leschziner said it would be helpful for all KLS patients (if they have not already done so) to send blood samples to Stanford for Dr Mignot's genetic research. To do so please contact Ling Lin at Stanford.

Dr Leschziner would like to collect blood samples of people with KLS in the UK between and during episodes to see the different chemicals in the blood. He is hoping to employ a travelling phlebotomist to take samples at home. We will update you when this research starts.

Dr Leschziner introduced Dr Valentina Gnoni who is doing research comparing migraine and KLS and is keen to extend the study. To be involved in this research you need to be a patient under the Sleep Disorders Centre at Guy's and St Thomas'. The research involves completing a questionnaire about migraine in family members and the KLS patient. **If you would like to take part in this research please contact Dr Valentina Gnoni.**

Q&A with Dr Leschziner

Is KLS an autoimmune condition?

Although steroids may work on the inflammation component of KLS this does not mean that KLS is an autoimmune disease. There is no clear evidence that steroids work and taking them is not without risk.

Is there any link with HPV Vaccination?

No

Do stimulants help at the tail end of an episode when the sleep is less pronounced?

KLS patients often experience anxiety and want a familiar space, watching familiar things on tv. Drugs to keep a patient awake will worsen anxiety.

KLS and comparison to other sleep disorders.

KLS is a 'Syndrome' which means there is an association of multiple symptoms rather than a disorder. There is significant variation between KLS patients.

Dr Laura Madeley

Dr Madeley's slides are available on the KLS Support UK website.

Dr Madeley spoke about the importance of the breakout session at the Family Day so people with KLS can speak together about their experiences and problems. The impact of KLS on a person's life is going to be different as everyone has a different KLS experience.

Dr Madeley reported on the findings of the Paris group on the psychological impact of KLS. KLS patients suffer slightly more anxiety than the general population. Other psychological difficulties such as depression are no higher than the general population.

Challenges of KLS

The challenges round KLS include the rarity, intermittent nature, range and efficacy of treatment, managing episodes, impact of episodes, managing time between episodes, age of onset and impact on life and how you discuss the diagnosis with others.

Diagnosis -getting an accurate diagnosis can be a challenge and can take a long time which creates uncertainty.

Treatment during bouts – As the advice is care at home and generally no medication this watchful non-intervention can cause anxiety

Age of onset – a challenge as it determines how you discuss the condition.

Psychological interventions

Create ‘formulation’ using 3P model –

Predisposing factors - you can’t change genetics

Precipitating factors – understand risk factors where possible. Some of the triggers may be able to be managed such as sleep deprivation, drug and alcohol use.

Perpetuation factors - anxiety is worse if people suffer long episodes.

Develop Management plan for episodes and coping after episodes

Adjustment to diagnosis - how to live as full a life as possible

Creating ‘safe uncertainty’ in remitting-relapsing conditions

Management of other co-morbid difficulties (if present) e.g. anxiety

Psychological approaches

Cognitive Behavioural Therapy

Acceptance and Commitment Therapy

Mindfulness Based Stress Reduction

Compassion Focused Therapy - build resilience to the difficult challenge

Where to access support?

Sleep disorders services, Improving Access to Psychological Therapies Services (IAPT) through GP or self-referral, CBT / Mindfulness based approaches, Universities / Colleges

Q&A with Laura

What do you do if the school counsellor does not understand KLS and makes matters worse?

School counsellors do not have specialist expertise but they should be able to help with anxiety and low mood. If you have had a bad experience of psychology and counselling she encouraged people to give it a second chance. She also stressed the importance to direct counsellors to the kls-support.org.uk website and KLS Support UK leaflet to help them understand KLS.

Tea, Break out session and Discussion

There was then tea and an opportunity for families to get to know each other and exchange experiences. This was followed by a break out session for the young people with KLS facilitated by Dr Madeley and an informal discussion for the families.

During the discussion some of the following points were discussed.

Benefits/support were being received by some families (in some cases after court hearings) - Education, Health Care Plan (EHCP), Personal Independence Payment (PIP), Young Sibling Carers support, Disabled Student allowance for university students, the Expert Patient Programme.

An advance directive or hospital passport is useful to give the parent/carer permission to talk on behalf of the person with KLS.

Carers are entitled to free flu vaccinations.

Research

In the UK

Jake Begley would like to thank all those who participated in his research on Exploring the Journey from Onset of Symptoms to Receiving a Diagnosis in KLS. We will share his findings once the paper is published.

KLS has been selected as one of the rare diseases to be researched by Genomics England. If you would like to take part in this important research, please let us know and we will put you in touch with the researchers. KLS is also part of the NIHR Bioresource and to take part in this please contact Dr Gerome Breen.

In the US

Dr Emanuel Mignot has a post-doc position open for KLS research. It is also open to post docs from outside the US. For more information.

If you have not already done so, you can take part in Dr Mignot's research.

Raising Awareness

As part of its aim to raise awareness of KLS amongst the medical profession KLS Support UK presented a talk on KLS to medical students studying sleep at Warwick University.

We would like to thank all those who have raised awareness of KLS in the media this year including Hermione Cox- Denning and Megan Firth. Raising awareness helps others find a diagnosis. Keeping KLS in the media also helps family, friends, teachers, work colleagues, the public and medical profession understand KLS. We are regularly contacted by journalists and producers interested in KLS. If you would like to be involved in future media projects, please let us know. We will be happy to support you through the process.

Fundraising

Thanks to all of you who have been involved in fundraising or have donated to KLS Support UK.

Rival Ride 17 -18 May 2019

A special thanks to Wasps Legends for their grant and support. Jok Dunbar is organising a bike ride from Harlequins home ground to Wasps home ground finishing in time for the last game of the rugby premiership. The ride will raise money for a number of charities including KLS Support UK. If you are interested in taking part in the ride and would like more details please email.

We would like to thank Debbie for running the Great South Run and raising £400. You can still support Debbie and donate.

Thank you also to Jake Renouf and the Renouf family for their continued fundraising for KLS Support UK. If you would like to donate to Jake's Man v Mountain Challenge.

If you are doing a fundraising event for KLS Support UK, please let us know and we will be happy to publicise your event through our Facebook page or website. If you would like us to help you in fundraising for KLS Support UK by providing a T shirt or setting up an online account through Mydonate please get in touch.

If you would like to donate to KLS Support UK we have a fundraising page on Mydonate.

If you would like to donate whilst shopping, it is possible to support us through Easyfundraising.

Funds raised are used by KLS Support UK to carry out its objectives of support, raising awareness and supporting research. We do not receive grants and the trustees are volunteers. Funds have been used to provide KLS Support UK meetings for families, raising awareness at medical conferences and producing leaflets, posters and T Shirts to raise awareness.

Support

We continue to provide support by telephone, email and messaging through the website. If you would like to talk to one of us please send us a message through the website or an email and we will call you.

We are also happy to put families in contact with one another if requested either personally or through our closed Facebook group.

You are receiving this Newsletter as you have agreed to receive emails from us. If you do not wish to continue to receive emails and newsletters from KLS Support UK you can UNSUBSCRIBE to remove your contact details.

Best Wishes

Adele Clarke, Jok Dunbar, Danielle Harris, Beverley Hunt, Caron Krieger

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