

Newsletter

Nov 2008



Friends of Landau Kleffner Syndrome
(Regd. Charity No. 1059499)

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Introduction

"Season's Greetings" to you all, as we enter the festive period. It's hard to believe that it has been six months since our last newsletter, wherever does the time go?

It still feels as though my entire wardrobe has been worn in every month this year as the weather continued to be most 'unpredictable', a word, which as a parent of a LKS child, we all know too well!

As I look back and reflect on 2008, I feel extremely proud of what we have achieved and as always, I find it a privilege to be the chairperson of FOLKS. I really do hope you find this newsletter of interest. We always aim to cover a wide range of topics and this newsletter is no exception.

On the fundraising front, I would like to acknowledge the fantastic support and awareness raised for LKS by the Kenny family and all our Irish friends, as a result of their "pooling" together fundraiser. (I really must visit all the bars just for the craic)!!!

In addition, our Gala fundraising dinner was attended by over 200 people and raised in excess of £10,000 – a HUGE thank you to all the committee members for their hard work in making this event a fantastic success and to our dedicated friends, colleagues and family's, the generosity was quite simply overwhelming. Events such as this allow us to support essential medical research into LKS and we were delighted to be able to present Professor Neville from GOSH with a cheque for £30,000 towards their LKS research project.

Christmas cards again can be ordered from our website, orders are being received daily which is encouraging, thank you to everybody that has ordered so far, and if you haven't yet purchased your Christmas cards, please make a point of ordering a pack or two from us. Log on to www.friendsoflks.com and purchase the cards you want using Paypal. Alternatively download the form, select the cards required and post along with your cheque to the address printed on the form. Either way your order will be sent without delay.

As we head towards the festive season, may I take this opportunity to wish you all a very Merry Christmas and a prosperous New Year (which may be a challenge with the current economic climate). However, as we all know, our health is life's most precious gift and I therefore wish you and yours prosperity on that front in 2009.

As a charity our objectives next year will be to hold a 'Parents Day' in Spring and by popular demand a 2nd Gala Charity Ball in Autumn. We will also continue to help families in anyway we can, in what are often challenging, difficult and worrying times.

Please remain positive, every cloud has a silver lining!

HAPPY READING

Angie Conlon

Member story

For most people, speech, the ability to listen and understand language is taken for granted. It is a mundane and frequently automatic part of our lives. But, what people forget, is that it can all change in an instant.

In March of 1992, I was a very happy healthy 7 year old. I was smart, outgoing and aside from a minor case of asthma, I had no health problems. Then, all of the sudden, I began to have great difficulties understanding the world around me. It was like I couldn't hear what people were saying to me. My parents would give me simple instructions like, "Elena, turn the TV off and come eat dinner!" and I would not respond. They just figured I was being deviant or rebellious as children can be at times; so they didn't think much of my behaviour. Then, my school teachers started noticing a change in me: I was no longer the curious and eager to learn child in the front of the class that I once was. One afternoon, my 3rd grade home teacher called my parents and told them that I was wondering around the classroom while she had been teaching as if I was bored with what she had to say or couldn't hear her. Within weeks, my school requested I have my hearing checked before my aloofness became any more of a disturbance at school. But my hearing was fine and all of the tests came back normal. So I returned to school.

Unfortunately, my problems only worsened. Not only was I having trouble understanding what people were saying to me but now I began to have problems speaking. I couldn't say what I meant and it was subconscious. I would say, "Mom, look at all the shoes in the sky," and then shake my head and say, "Oops, I mean birds". I would switch up words in sentences or eliminate certain words all together. This is when my parents really knew I had a problem. I stopped pronouncing long words correctly and even began confusing letter sounds as though I was a 2 year old learning my ABCs and how to read all over again. One of the worst parts of being 'sick', as I was, is the negative bias that comes with the symptoms. Because I eventually lost most of my verbal communication skills and had to have people speak very slowly and use visuals to communicate with me, I was classified as 'retarded' in my school. My principle held a meeting with my parents, along with the district education board and insisted that I needed special education that his school could not provide for me. He assumed I was mentally retarded or autistic and that I did not belong with normal students.

This is where my parents' hope, persistence and diligence was vital. They were determined to find out what was wrong with me and help me get through it. I started going to different doctors. First, they did think I was autistic but I had had no prior developmental abnormalities. There had been no delays in my motor and language development (as was typically noticed in children with autism) until

spring of 1992 when I was at the age of 7. I had no illnesses or accidents that could have caused central nervous system damage-that could have affected my speech and language usage. I had normal audiology evaluations and my head MRI scan came out completely normal. Doctors were puzzled and I just kept getting worse; my mother even considered home schooling me. Finally, in late August, a doctor from Scottish Rite Children's Hospital noticed that I had major spikes or epileptiform discharges in the left parietal-temporal area of my brain from the electroencephalograms (EEG) he had taken. I was basically having mini-seizures all the time in my brain which was preventing the neurons and information transmitters in my brain from working correctly. But the seizures I was having were not like the ones associated with epilepsy; my seizures would occur very frequently, for 30 seconds to 2 minutes and I would just have this blank stare on my face and not respond to anything around me. The doctor diagnosed me with acquired epileptic aphasia, which is also known as Landau Kleffner Syndrome. My parents couldn't understand why this was happening to me because we had never had any kind of learning disabilities, seizure disorders or neurological conditions in our family history. To add to their distress, the doctor told them there were so few prior cases, that they would just have to experiment with different dosages of medications to see if I would improve.

I was first prescribed Klonopin, which made me very sensitive (usually aggressive and irritable for no reason) and I was crying all the time. Then, I was put on Depakote, which showed very good improvement for the first couple of weeks but then gradually stopped working. The doctors tried different variations of Prednisone (which gave me nose bleeds) and Phenobarbital, but they both had negative effects on my recovery. There was another doctor who actually suggested I go to UCLA and have an experimental surgical procedure done where they would drill holes in my brain and it 'may cure' my condition. I'm so glad my parents never let that happen. Eventually, through much trial and error, the doctors found the right combination of medication for me: Tegretol and Klonopin. My parents said as soon as I started taking that, they could see I was finally showing signs of improvement. In addition to the medication, extensive dual therapy between my own private speech therapist and the two that the school provided for me also greatly helped my improvement. They helped me rebuild my confidence in my speech as well as my self-esteem, after being harshly bullied and labeled at school.

By February of 1994, six months after my 9th birthday, I had almost completely recovered. I regained my vocabulary, understood people when I was being spoken to and was back to my normal

outgoing self. Before, people had such a hard time understanding me because my speech had deteriorated so much that I became so frustrated, I just chose not to speak anymore. I was so glad to have people understand me again. I was lucky because my parents really fought to have the district let me stay in a normal public school. One of their biggest arguments to the board was that LKS, ironically enough, never affected my IQ. I had always tested in the top 2%; and despite my altered language and speech skills, I still excelled in arithmetic, calculations and visual/motor tests.

This all happened in the pre-Google age where the internet was still new and people still had to do research the old fashion way. My mother spent many hours in public libraries, shifting through confusing medical books, trying to find the answers to LKS. Fortunately today, parents whose children

are diagnosed with LKS have a lot more information available to them than my parents ever did. There have been more cases, more research and most importantly, doctors with more experience with the syndrome. Plus, with websites like www.friedsoflks.com, parents have support and realize they're not alone.

Today, I am 24 years old college graduate and am blessed to have LKS behind me. I am forever grateful to my parents for never giving up on me and the doctors and speech therapist who believed in my recovery. I appreciate life and I know not to take things for granted for it can all change in an instant.

Elena Karenev

LKS and Auditory Processing Disorder

Linda Philips is a speech and language therapist who worked with children who have LKS, at Great Ormond Street, for some years. She undertook a research project looking at LKS and auditory processing disorder. This is an account of her study, and she would like to thank Dr Imogen Newsom-Davis and Dr Tony Sirimanna, for assistance with data collection, and Paul Turner, for supervision.

In addition to language difficulties, children with LKS often experience a range of problems related to the processing of sound information. These difficulties may include:

- short term memory
- hearing the differences between similar sounds
- following spoken language in real time
- coping with background noise or situations where there are multiple speakers.

By contrast, children with LKS often benefit from information presented visually or where modifications to spoken language are made (e.g. slowing down, repetition and reducing background noise). Difficulties of this nature have been described as an auditory processing disorder (APD). There is, however, considerable controversy as to whether this term applies in the case of LKS or not (Stefanatos 2002, Korkman et al 1999).

Individuals with APD typically have normal hearing but experience problems with the way that auditory information is **processed** in the brain. The American Speech and Hearing Association (ASHA) define APD as: "a deficit in neural processing that is not due to higher order language, cognitive or related factors". ASHA (2005) recognise that APD may exist alongside other disorders but make the proviso that it does not occur as a **result** of these disorders.

There are a number of reasons for the debate surrounding LKS and APD. Some have argued that the nature of the language disorder and the fact that the problems cannot always be fully circumvented by by-passing the auditory channel is evidence against a diagnosis of APD (Korkman et al 1999). Research findings may also differ depending on when the assessment is conducted, with the expectation being that APD-type difficulties are more likely to occur earlier on in the process. Added to this, is the controversy surrounding the definition of APD which has only recently been clarified. Researchers (Institute of Hearing Research) in the field are currently developing a comprehensive test battery which should reduce confusion. Finally, research on LKS typically involves single cases or small groups, which restricts the potential to determine common trends. The relevance of this theoretical debate lies in the need for professionals to provide the best possible therapeutic and educational management programs for children with LKS. As the intervention programs differ depending on the nature of the disorder (i.e. language versus auditory based problem), it is important to try and resolve this issue.

In an attempt to address the above, research was carried out with the following questions in mind:

1. Does APD exist in late outcome of LKS?
2. Are there any particular aspects of APD that are universally impaired?
3. Are there any specific language or cognitive deficits that can be observed?
4. Are there any behaviours that are common to everyday life / learning experiences?

Four participants (11.6 to 15.7 years) who had

attended the Developmental Epilepsy Clinic at Great Ormond Street Hospital were invited. Each participant had a full language, cognitive and audio logical assessment and also completed a range of questionnaires considering their behavioural profiles.

The results showed that all participants had problems with auditory processing, with processing of time related information being notably implicated. Although a causal relationship between APD and LKS cannot be established, the evidence supports the co-existence of APD and language / cognitive difficulties. Despite participants being selected for their relatively good outcome, language scores were compromised in three out of four individuals and phonological processing (processing of speech) was universally impaired. The cognitive assessments revealed variable results affecting either or both verbal and performance skills. There was strong indication of short term auditory memory difficulties. On the questionnaires, emotional symptoms and hyperactivity were frequently reported by either parent or child and to a lesser degree, problems with peer relationships. Notably, teachers did not tend to recognise these issues which suggest that they are complex and subtle in nature.

The existence of APD in the late outcome of LKS, strongly suggests that these features would be more pronounced earlier on in the course of the disease process (given the presentation at onset). Overall, the results confirm the need for a highly comprehensive multidisciplinary assessment. In addition to neurology and speech / language

therapy, there is a need for audio logical assessments to be added to the current battery routinely employed. The routine involvement of a psychologist should also be strongly considered to ensure both suitable educational placements and to address behavioural / emotional issues where they arise.

REFERENCES

- American Speech-Language-Hearing Association (2005) (Central) Auditory Processing Disorders
<http://www.asha.org/members/deskrefjournals/deskref/default>
- Institute of Hearing Research, Medical Research Council website:
<http://www.ihr.mrc.ac.uk/research/projects/apd/imap.php>
- Korkman M, Granstrom M, Appelqvist K, Liukkonen E (1999) Neuropsychological Characteristics of Five children with the Landau-Kleffner Syndrome: Dissociation of Auditory and Phonological Discrimination *Journal of International Neuropsychological Society* 46 (6) 566-75
- Stefanatos GA, Kinsbourne M, Wasserstein, J (2002) Acquired Epileptiform Aphasia: A Dimensional View Of Landau-Kleffner Syndrome and the Relation to Regressive Autistic Spectrum Disorders *Child Neuropsychology* 8(3) 195-228

Research News

Landau-Kleffner syndrome is a devastating condition that robs a child of language and can also cause problems with social interaction, attention, reasoning and even movement. It is rare and we understand very little about

- why certain children are affected
- what is happening to the brain to cause the loss of skills (we know that it is associated with active epileptic activity, but not how this affects brain development and function)
- why there can be such circumscribed loss of abilities, for example sometimes losing the ability to understand spoken language but retaining the ability to understand other sounds and to read
- what is the optimum treatment
- why there can be such a wide range of outcome, with some children recovering fully, and others suffering lasting impairments

At Great Ormond Street, we have been working with affected children for more than 15 years, which means that we have seen well over a hundred children with LKS. Over that time we have done some research studies to develop assessment tools, establish the effectiveness of corticosteroids, evaluate use of gestural communication and examine auditory processing in recovered cases. Over the same time, our clinical approach has evolved with early and carefully monitored medication, establishment of pulsed (weekly) steroids to maintain any improvements, careful assessment for surgical suitability and close liaison with local health and education teams to support the child.

To date, we have been primarily a clinical service, but with FOLKS help we intend to take a lead in research. FOLKS have kindly donated £30,000 and we are going to use this money for basic scientific research into LKS. The neurosciences unit at GOS is already actively involved in investigating the origins and outcome of other forms of epilepsy and we will be able to build on their expertise. One team under Professor Neville has been

investigating infantile spasms, which is an early onset epilepsy that often has catastrophic and lasting effects on a child's cognitive abilities, not dissimilar to, but more pervasive than LKS. They used a neurophysiological investigation called event related potentials (ERP's) to assess the developing pathways for processing complex sounds and showed abnormal responses, giving insight into how epilepsy can disturb early brain development. We intend to use a similar technique to investigate brain function in LKS.

At present, we understand very little about the fundamental disturbance in LKS. Of course we observe its effects through the child's acquired inability to understand speech (receptive aphasia), but we do not know whether the basic difficulty is with processing language, or processing speech sounds, or both, or a related brain function. We hope that ERP's will clarify this, and may even discriminate whether a child will respond to treatment and prognosis.

Over the next year, we will focus the study on 2 groups, children who are currently attending clinic with active disease (including their response to treatment) and those who no longer have active disease (both good and poor outcome). This latter group may mean that people are invited back for a special review.

In the developmental epilepsy clinic we are often asked by parents and young people, what the future holds after LKS and what life as an adult will be like. Currently, it is very difficult to predict outcome, as it depends on the particular child's skill profile, the disease process (age of onset, number and severity of regressions, length of active disease, response to treatment), and progress in different skill areas over time. Until now, outcome research has tended to focus on language skills. LKS can be conceptualised as primarily a language disorder, but other cognitive functions are also involved including attention and organisation. These skills are increasingly important, as the young person enters adolescence and more independent thought and behaviour is required. Other factors such as emotional development and social-communication will also affect the person, their families and people in their social networks. However comprehensive exploration of these dimensions has been neglected in the literature. We will therefore combine the ERP project with a cross sectional study, looking at how children who have had LKS in the past, are functioning now. This will help us to have a broader and more detailed understanding of outcome, and hopefully give insight into appropriate provision for these children and young people.

Recently, certain epilepsies have been shown to have a genetic basis. Given our limited understanding of LKS, it would seem reasonable to consider genetic mechanisms. There is a joint research group from the Institute of Child Health

and the Institute of Neurology that is able to process blood samples for us, looking for candidate genes so we propose adding this to the research, particularly as samples will be stored for future advances.

As you can see, we are hugely ambitious about the research and are committed to it as a team. FOLKS money will allow us to employ a full time researcher, in addition to our official clinical commitment of 3-4 clinics a month. It may be best to regard this year as the first step towards establishing a larger programme of research into LKS. In the future we would like to develop a study of brain imaging (MRI) at various stages of the natural history looking for loss of brain tissue secondary to subclinical seizure activity. We also hope that the results from this study would allow an application for further research funding as well as suggesting new avenues to pursue.

LKS is one of the conditions being studied as a part of a new European Community Grant on rare disorders. This will entail a data base within the European partners and be particularly concerned with how to deliver high quality care to families and children with rare disorders. Our centre is designated to take a lead in this.

Summary of current project:

We will invite 20-30 children and young people to enrol in the project. The study will be registered at Great Ormond Street Hospital and informed consent taken. For current patients, it is likely to add 2 hours to the routine appointment and involve some questionnaires that can be filled in beforehand. For others, it may take 1-2 days, particularly if we do not have recent or stable neuropsychological data. The assessment will include:

- **Auditory ERP's:** This takes place in a sound attenuated booth, with the parent outside the booth, but able to see the child via a closed-circuit monitor/ glass window. Before the ERP session, electrodes, rather like those for an EEG, are placed on the scalp while the child is distracted (e.g. with suitable videos). The actual ERP session takes about 30-40 minutes and involves the child viewing pictures on a monitor and listening to sounds or words through headphones, which may or may not match the pictures on the screen. The child does not need to respond to or remember the pictures or words.
- **Neuropsychological and language** assessment (unless recent/stable profile exists)
- **Social and Quality of Life** profile
- **Blood sample** for genetics study

Team at GOSH:



The Developmental Epilepsy Team first started in 1989 through the determination of Brian Neville to offer a holistic service to children with complex epilepsy. It is a truly multidisciplinary team that includes a speech and language therapist (**Nicola Jolleff**), a clinical psychologist (**Monique Cloherty**) and neurologists (**Brian Neville** and **Maria Clark**). The FOLKS research donation will allow **Michael Kihara** to join the team and carry out ERP's and assessments. Michael is a psychologist who has been working for a research team in Kenya (Kilifi) that partners with GOS. He is well known to us and has extensive experience with ERP's in children with brain problems, including epilepsy.

Contact details:

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Gala Fundraising Dinner

On Saturday 27th September FOLKS held its first Charity Gala Dinner at the Heathrow Marriot in London to raise funds for research. A very successful and highly enjoyable evening was attended by over 200 guests.

On arrival guests were treated to a champagne reception before being ushered into the main hall by Master of Ceremonies John Willmott. After being served a delicious three course dinner John introduced FOLKS Secretary Richard Budnyj to the stage to say a few words about FOLKS, its fundraising and the purpose of the evening ending with a thank you to all those that had attended. Richard then introduced a very brave Michael Conlon aged 12 to the stage to tell in his own words how LKS had affected him and why people should support FOLKS. He then presented a cheque for £30,000 from FOLKS to Professor Brian Neville to be used to kick start the research programme. After receiving a standing ovation Michael then handed back to John who kicked off the evening's entertainment.

Firstly a very successful auction with all items going for more than the minimum bid value set. Next up was Sara Lawson who sang three songs from popular musicals before the dance floor filled up with the musical talents of the Squealing Pigs who rocked the night away.



During the night activities such as raffles, number boards and Heads and Tails help fill the coffers. In total the evening over £10,000 which far exceeded our expectations. So all in all a very successful and highly enjoyable evening one which we hope to revisit next year.

Finally we must thank all those people that made the evening possible and contributed to a successful function. Firstly Angie, Marie-Suzanne and Cathy whose attention to detail meant everything ran smoothly. To the Marriot hotel whose service and quality was outstanding. To all the entertainers, Sara Lawson, the Squealing Pigs, the table magicians and not forgetting John Willmott in all his outfits who was an excellent MC. To Everton Stephenson for the photography and also the lovely Bath students who helped on the night to persuade guests to give up their money. To

all those kind donators of auction items, raffle prizes and money towards the events and finally all the guests who supported the evening and

contributed to raising such a great sum. Thank you to all. See you next year!

'Pooling' together Fundraiser

During the August Bank Holiday Weekend, members of the Manchester Pool League once again travelled to Ballyshannon, Co. Donegal, Ireland to play a pool tournament against teams in Ballyshannon and surrounding areas. Following on from last year our second trip was as successful with just over £3400 raised for FOLKS. The trip was organised by the Kenny family in Manchester, whose nephew, James, was diagnosed with LKS several years ago.



The Manchester team played the Irish teams in various venues throughout the weekend and we

would like to say a big thank you to the proprietors of the Black Cat (Belleek), the Travellers Rest (Cashellard), Sean Ogs and Macs Bar (Ballyshannon) for making us so very welcome and helping us to raise much needed funds for a charity so close to our hearts. The generosity and warmth of the Irish people was again outstanding.

A big thank you to Brendan Gallagher, Ballyshannon who worked very hard on the Irish side to ensure the weekend went smoothly. We really couldn't have done it without him.

Following the events of the weekend, a local paper ran a story on the trip, describing LKS and highlighting the efforts of FOLKS. We found out later that there is a child living near Ballyshannon who has LKS. Both families have since been in touch. This just highlights further the good work that raising awareness does.

Needless to say a great time was had by all and we are looking forward to doing it all again next year!

**Ros Kenny
Manchester**

And Finally.....

Firstly and once again a big thank you to all the contributors of this newsletter and to all those that have raised money for FOLKS over the last year. It is fundraising and donations that allow us to continue as a successful charity. So keep thinking and acting on ways, in which, we can raise more funds to support our goals for the future. Be it £50 or £5000 it all makes a difference so think what you, a relative or a friend could do in 2008.

Secondly, our big event this year was as you read above our Gala Charity Dinner. As I said in the last newsletter we were very disappointed to cancel the last family day but we will try again in Spring 2009. But remember it is for the members benefit so please make an effort to attend the next get together. Giving the opportunity to get together once a year is a real benefit to those struggling to deal with this condition. I know as a parent of a child with LKS and also as a trustee of this charity that sharing our experiences is invaluable and we are all more than happy to do it, as we relied on the

support we received in the early years. So if you are new to FOLKS and struggling to understand LKS or a seasoned LKS parent then please take advantage of the opportunity.

And finally as I have stated many times in previous newsletters we would gratefully welcome new blood to the committee. I make no apologies for continually reminding everybody but without new blood we don't move forward. We meet up every other month for a couple of hours. If you don't feel you can join the committee but still wish to help FOLKS in some other way please contact us 0870 8470707 or e-mail us at info@friendsoflks.com.

Many thanks and I look forward to your continued support.

Have a great Christmas

Richard Budnyj

Secretary

F.O.L.K.S.NEWS welcomes all contributions, articles, letters and comments for publication. If you have any item suitable for publication it should be sent to Richard Budnyj, Secretary, 8 Malibres Road, Chandlers Ford, Hants SO53 5DT, alternatively e-mail to info@friendsoflks.com. F.O.L.K.S. does however reserve the right to omit or edit items where necessary. F.O.L.K.S. NEWS is published to provide general information to parents and carers of children with Landau Kleffner Syndrome and to interested professionals. The contents are not and are not intended to be, a substitute for advice from a qualified medical practitioner, preferably one experienced in the management of this complex disorder.

Executive Committee:

Angie Conlon (**Chairperson**), Richard Budnyj (**Secretary**), Steve King (**Treasurer**), Cathy Cowie, Martin Cowie, John Conlon, Robert Duncombe, Patrick Magee, Marie Magee.