



Great Ormond Street Hospital for Children NHS Trust: Information for Families

# Landau Kleffner syndrome: abilities, behaviour and education

This information sheet is part of our series on Landau Kleffner syndrome (LKS) and focuses on abilities, behaviour and education. Other topics covered in the series include: an overview of Landau Kleffner syndrome, language and communication and what you can expect from Great Ormond Street Hospital (GOSH).

## Contents

Abilities	1
Behaviour	3
School	7
Further information and support	12

## Abilities

### Language

This is covered in our *Landau Kleffner syndrome (LKS): language and communication* information sheet, available from the team or on our website.

### Non-verbal skills

Everyone has abilities that help us to understand the world around us, often called non-verbal or performance skills. In children, they include skills such as visual matching, drawing, design and construction, geometry and mathematical problem solving. In general, these abilities are not severely affected in children with LKS. This is important, as the child can use these skills to support their learning, compensate for their language difficulties and boost their self-esteem.

In a minority of children, non-verbal and language skills are affected equally, so there is a 'global' pattern of difficulties across all areas of learning and development. This is more common if LKS develops at a very early age.

### Memory

Although it may only be apparent in the older and more able child, LKS can affect memory, especially for verbal material. Careful assessment is necessary to distinguish between problems due to retrieval (getting information out) rather than difficulty encoding (getting information in). Difficulties with attending to and/or processing incoming information may also lead to memory difficulties as the child has not been able to process the information in the first place.



Memory difficulties are a direct consequence of the abnormal brain functioning that occurs in LKS. A variety of strategies can be used at home and at school to minimise the consequences of poor memory. These include using simple visual mnemonics (memory prompts such as 'Richard of York gave battle in vain' to remember the colours of the rainbow), timetables, checklists of what to take to school, reminders on mobile phones and so on.

### **Executive function**

These are the cognitive processes that allow us to organise our thoughts, feelings and behaviour, anticipating outcomes and adapting to changing situations. They include the ability to plan, start and monitor actions, stop inappropriate actions and select the information on which to focus. These skills develop in middle to late childhood and can be affected by LKS. It is not uncommon for difficulties to become more apparent as the child grows older, particularly as they move on to secondary school.

### **Motor skills**

Around two-thirds of children with LKS have problems with movement, particularly during the active phase of the condition. Difficulties may include dyspraxia or lack of coordination, shakiness or tremor, unsteadiness, jerky movements, unusual limb postures, weakness or even neglect of one side of the body. This may affect activities such as writing, dressing, walking and may make it difficult to use gestures or signing. The muscles around the mouth and throat are commonly involved and may cause difficulties with feeding, controlling saliva and speech.

In certain cases, the child may experience weakness following a clinical seizure or sometimes loss of speech. These immediate post-seizure difficulties usually get better over a few hours or occasionally days. However, some children change hand preference following this type of episode.

In general, these difficulties with movement are among the first areas to show improvement as the child recovers.



## Behaviour

At least half of children with LKS experience neuropsychological and behavioural difficulties and the most common types are described in this section.

### Attention deficit, hyperactivity and aggression

Many children with LKS have poor attention or over-activity, often associated with irritability and aggression directed towards a particular family member. These features may be **mild**, where the child is a bit more 'bouncy' than usual, has become slightly impulsive or has difficulty keeping concentration for an entire lesson. They could also be **intermittent**, for example a couple of over-active hours in the evening or at large gatherings with a lot of noise and stimulation, or **severe**, such as affecting all activities consistently and having a diagnosis of attention deficit hyperactivity disorder (ADHD).

The most common features reported are inattention, hyperactivity, impulsiveness (not thinking before doing or saying something), no sense of danger, verbal and/or physical aggression, mood changes and lack of inhibition (failure to control inappropriate behaviour). These behaviours are due to the disease process, which means that the child probably has very little control over these aspects of their behaviour. They are not simply a response to the frustration and confusion felt by the child at the loss of language.

Often, ADHD-type behaviours improve if the disease activity is controlled. When severe or persistent, it is important to treat these ADHD-like difficulties in their own right, as they may prevent the child using other skills to learn and interact. It is often most effective to use a combined approach through a behaviour programme and medication such as methylphenidate (Ritalin®).

Although these behaviours are due to the disease rather than any conscious control by the child, they are managed through behavioural techniques. These children cannot learn through oral communication, such as instructing, warning or praise, so alternative ways to teach such behavioural control is necessary. There may also be a 'learnt' component as these behaviours may produce a desired outcome, for example if a child has a tantrum and throws things around when the television is turned off, then someone will turn it on again. This means that the behaviour will occur more frequently as it is 'rewarded' by the consequence. It is important that parents should be aware of this possibility and stick firmly to their pre-determined rules where possible and continue to provide as calm and structured an environment as possible. Although allowances must be made because of the involuntary nature of some of these behaviours, it is still important to make clear what is and is not acceptable and to develop strategies to deal with common situations.



Useful approaches include:

- immediate and consistent responses to behaviour
- time out
- distraction techniques
- rewards for positive behaviour and achievements

The advice and input of a local clinical psychologist, often from the Child and Adolescent Mental Health Service (CAMHS), may be necessary to help resolve situations where behaviours have become very challenging. It is usually helpful to discuss these matters openly with the school, so that appropriate boundaries and responses to the behaviour can be agreed to ensure a consistent approach.

In children with milder difficulties involving 'cognitive' inattention and impulsivity, the following strategies may help:

- playing games that require attention and memory to encourage these skills, ensuring that the games are at an appropriate level of difficulty so that the child experiences achievement rather than failure – try the Early Learning Centre® for suitable games
- the parent counting to ten before responding to a situation that is upsetting
- discussing basic rules to help with impulsivity, such as 'stop and think'
- creating simple visual mnemonics (memory prompts) to help remember important verbal information

## Sleep disorders

As LKS is particularly associated with seizure activity during sleep, it is perhaps not surprising that many children have problems at night. Often they cannot settle to sleep until late or they may have prolonged episodes of wakefulness during the night or be woken by seizures. Also some medications, such as lamotrigine, may disturb sleep. Children who have difficulty getting off to sleep may be helped by melatonin (see information sheet for details). It is harder to treat night time waking but the situation can usually be improved by consistent use of standard behavioural management strategies, including:

- a regular, quiet bedtime routine, such as a bath, warm drink, reading or looking at a picture book
- removing televisions and videos from the child's bedroom
- sleep in their own bed in their own room, with a baby monitor if you are concerned you will not hear them when asleep
- comfort and reassurance on waking without making a fuss, for example resisting switching on lights, giving food, putting on a video or staying with the child until asleep



### **Other behaviours**

Some children with LKS become very tearful and/or depressed and this should be monitored carefully. They may require more reassurance than usual and become anxious in social situations. Others may become more controlling of their environment.

A small number of children become passive and apathetic in their manner. This is most commonly associated with a marked global regression and early onset before 2½ years of age.

Some children are extremely irritable and aggressive with violent manic outbursts. Others may develop obsessional behaviour, anxiety or severe impulsivity. They need psychiatric review and a few will require medication.

### **General support principles**

Language is the easiest and quickest way for most of us to communicate, find out information and record ideas. We do this through speech, reading and writing. Of course, it is not the only way. People also use facial expressions, gestures, symbols and so on. However, for most of us, language is fundamental to how we live. For the child with LKS, the effect on language may be such that the world remains familiar but is transformed so that people use a language that you cannot understand or speak. You might try to guess what is happening from clues around you, but it will be very tiring and unrewarding.

As language is fundamental to so much of what we do, the child with LKS needs a comprehensive programme to support them throughout the day, at home and at school. This is most effectively achieved if everyone is committed to strategies that help communication for the child. These strategies will vary with the child and disease severity, but will include common themes such as simplifying language and the listening environment, offering alternative communication strategies and providing visual reinforcement. The family provides the main care for the child and parents are the main communication partners. They should be actively involved in decisions and given appropriate information and support, including opportunities to learn specialist skills, such as signing systems, which can be used at home.

Children who lose the ability to understand environmental noise will need special support and supervision. Certain situations will be more dangerous for them as, for example, they cannot detect traffic noise or warning shouts. They may find crowded environments and group situations distressing, as they no longer have an auditory forewarning of what is about to happen or what is expected of them. This can also be true for children who retain some language but find it difficult to pick out speech in a noisy environment. Even playing team games such as football, where team members signal to each other verbally, can be difficult.



Some children become very sensitive to and intolerant of certain noises or even music. This is probably due to the brain processing the sound in an unusual way, such that it is perceived as an unpleasant stimulus. This may restrict family outings, as certain noises such as public address announcements, can be very distressing for the child.

In addition to language, the child with LKS often experiences difficulties in other areas, such as behaviour, motor skills and non-verbal understanding. These must be tackled with an integrated approach that supports the child in all environments. Therefore, the local team must be able to draw on a wide range of services and skills, including language therapy, psychology, psychiatry, physiotherapy, occupational therapy and social work for instance, in order to provide an appropriately tailored programme.



## School

School provides a vital framework for a child's recovery and management. It is the key medium through which teachers and therapists can support the child's learning and help him or her make sense of the world, as well as providing a stable social structure. Given the complex and unusual nature of the learning difficulties associated with LKS and the behavioural problems that may also be present, identifying a suitable educational placement can be difficult and will depend on the individual pattern of abilities and difficulties in each child and the ability of the school to meet these needs.

In children who show a good recovery, mainstream education may be the most appropriate placement. For other children who show a moderate degree of recovery, mainstream schooling may be possible with help from a learning support assistant (LSA) to provide a semi-adapted curriculum that is appropriate to the child's levels of ability. For other children who have more specific needs, it may be necessary to consider alternative settings to ensure a 'whole school' approach to the child's particular needs.

Children with a profound language loss will usually benefit from learning sign language along with their families. They may be well accommodated in language units where there is specific expertise in dealing with children with language disorders, although it is important to check the unit's particular focus and provision. Others may be more

appropriately educated in schools or units for children with hearing impairment.

Where more general learning difficulties exist, schools that cater for a slower pace of learning overall may be the best option. Finally, those with pervasive development disorders or autistic spectrum disorders may be best placed in schools or units catering for children with autism.

### Statement of educational needs

Children with educational needs are often first identified and placed on the School Action or School Action Plus level of the Code of Practice. If these levels of support are insufficient to meet the child's needs, a Statement of Special Educational Needs may be required. The statementing process is carried out by your local education authority and may take several months, involving assessments by local educational psychologists and speech/language therapists. It should automatically be reviewed annually although a parent or school can ask for a review to be brought forward if there is a marked alteration in circumstances, such as a regression. The statement will set out your child's current level of ability and highlight the key areas of difficulty, both in terms of abilities and behaviour, recommending what level of support or input is required to help them progress. Each school has a nominated special educational needs coordinator (SENCO) who should then take responsibility for carrying out the recommendations. This should include careful planning and drawing up of an Individual Education Plan (IEP), specifying the ways in which



your child's learning will be supported and teaching methods adapted.

**Please note:** If your child is being educated in the private system, the way in which provision is delivered may be very different.

### **Common educational issues**

**Fluctuation in abilities** – LKS can change rapidly over time, making progress at school erratic and unpredictable. Regular monitoring and updating of therapeutic and educational plans is necessary to respond to this. When the child's disease is active, performance can vary even within a day, making them susceptible to fatigue and difficulties in concentration. Teachers and LSAs must be made aware of this and careful timetabling of lessons may help to minimise the impact.

**Recognising and accessing preserved skills** – Despite having significant language difficulties, many children with LKS retain average or above average non-verbal abilities. However, most classroom teaching is invariably verbal. This means that a specialist teaching approach must be devised. It is vital that these good skills are recognised and that it is not assumed that the child has general learning difficulties simply because of the language difficulties. Visual processing is usually relatively spared and can therefore be used to compensate for problems in processing auditory information and as an alternative means of communication. Ultimately, these non-verbal skills may be the way the child compensates for any residual deficits and is able to function in later life.

**Behaviour** – LKS is associated with a number of behavioural difficulties that may be very disruptive to learning and school life, for example, poor attention and concentration, social communication problems or aggressive outbursts. It may be important to allocate resource to activities that are not obviously educational, but which are impairing a child's function significantly. For example, the child who finds social interaction difficult may need additional help in unstructured situations such as the playground. Other children would benefit from help to tackle behavioural problems that might otherwise take them out of the learning environment.

### **Common elements for success**

School placement is more likely to be successful given:

- a comprehensive and flexible approach giving appropriately targeted support throughout the day
- good communication between parents and school in order to make the most of any new developments in the child and achieve consistency in management of any difficulties
- regular monitoring of the child's abilities by specialists and effective dissemination of this information and related recommendations or strategies to all involved with the child
- teachers and LSAs who are motivated to learn about LKS, are sensitive to changes in the child and flexible in their responses to this, and can consistently implement suggestions from parents and therapists to maximise the academic





and social potential of the child

- an appropriate peer group, that is, a group of children with similar skills, difficulties or interests, that can provide a social network and friendship
- education of the child's peers so that they can have some understanding of specific difficulties and appropriate behaviour and responses, such as a 'buddy' scheme to help support the child

### **Secondary school issues**

The active, seizure phase of LKS tends to stop around this age and there is no longer the extreme fluctuation in abilities. However, in this 'burnt out' phase of LKS, the residual difficulties become apparent and the young person continues to have special educational needs, often with an unusual learning profile. They may recover good language and general abilities, but often lack the early skills that would normally have been acquired around the time that the LKS was active. They need explicit, targeted teaching of these early concepts, including grammar, maths, specialist vocabulary, two-way conversation skills and so on, before they are able to perform at the level of their underlying abilities.

These young people often have ongoing difficulty processing (making sense of) language around them, particularly in noisy environments, making it hard for them to function in a mainstream classroom without intensive assistance. This is often so severe that it is as if they have a hearing impairment, and indeed some complain that they cannot hear when they transfer to secondary

school even though formal hearing tests are normal. Sometimes this difficulty is due to very slow auditory processing of language, although the ultimate comprehension of the material is correct. This slow processing may prevent them making sense of everyday conversation or following oral classroom directions in real time, which will in turn impact on friendship groups and schoolwork. These young people are usually very aware of their difficulty with following language, but are often embarrassed and do not like to ask for help or repetition. Sometimes their expressive language abilities are in advance of their comprehension skills, giving a false impression of language competence. This can make it very difficult for school staff to notice when the young person is struggling.

Young people who had LKS as children often have an impaired working memory, particularly for verbal material, so that they cannot hold and manipulate information in the mind over short periods of time. This leads to failure to follow instructions, difficulties with activities that combine storage and processing of information, such as mental arithmetic or reading comprehension, and problems with place keeping, including keeping track of conversations. Difficulty with executive function due to LKS may also become apparent during teenage years, leading to problems with initiation, problem solving, attention control and behaviour.

There are often problems with low self-esteem and low mood. Transition to



secondary school can be very stressful as the young person loses previously-established support and friendship and needs to rebuild these in an environment that will have no memory of their earlier profound illness. It is also a time when they will be expected, and will want to, become independent and it is a delicate balance to provide the necessary support.

It is important that both the young person and the school tackle these issues and there are a number of common and helpful strategies, many of which were outlined earlier in this information sheet. We would also strongly recommend continued access to speech and language therapy during secondary school to support learning strategies as needed.

### **General support principles**

- In general, background noise or other distractions should be kept to a minimum.
- Verbal communication needs to be short, clear and simple.
- Information must be explained to the young person in a format that is understandable to them and that it tailored to their level of language development.
- It is essential that visual cues and supports are provided.
- They must be given plenty of time to process oral information and then check that they have understood, by asking "Do you understand?" and "Tell me what it is you have to do". If they do not seem to understand, repeat what you have said or rephrase it, using

different simpler words.

- They will also benefit from being given plenty of time to put their thoughts into words.
- Vocabulary and concept words needed for particular subjects, such as science, history, geography and maths, for instance, are best taught in advance so that the young person can effectively access these lessons.
- Encourage the young person to recognise any memory failure or difficulty with understanding and seek repetition or clarification. They should also ask the speaker to slow down if they are speaking too quickly.
- Those with working memory difficulties should repeat aloud things that they have to remember, such as phone numbers, lists or name, for instance.
- Using memory prompts, such as sticky notes, task planners or mind mapping can also be helpful.
- Young people may also benefit from help in breaking down complex or novel tasks into smaller components to help them to learn problem solving methods, as well as help with planning and organisation.

### **Specific patterns of impairment**

#### **Good non-verbal skills in conjunction with language impairments**

– Pictorial and symbolic cues can be used to back up or replace verbal explanations. It may also be necessary to adapt heavily language-based subjects, such as in English lessons. It is worth noting that, although number concepts are generally considered to be non-verbal, mental arithmetic is a verbal



skill and relies on memory so may be difficult for children with LKS. An additional unusual feature affecting some children with LKS is that spelling and writing skills that have already been acquired may be retained during an episode of regression, so that the child may still be able to write and spell words that they are not able to understand or produce in speech.

**Impairment of verbal memory and auditory processing** – Slow processing of information and impaired verbal memory make it even harder for children with LKS to use their language skills. For example, they may understand language in a one-to-one session but in a noisy classroom be unable to decipher the same auditory information. In other instances, the child may understand spoken information at a simple level, but have auditory memory problems that mean that they are unable to remember a sequence of verbal instructions or a story, which could cause enormous difficulty in class and with playmates.

Where the child has retained a reasonable level of language comprehension, the following will usually be helpful:

- repetition of verbal instructions
- preferential seating, close to the class teachers
- reducing speech rate
- reduce background noise and distractions
- short and simple written (or symbolic) forms of communication where possible
- break work down into small chunks
- allow longer for the child to respond to questions

- lower expectations for subjects that are very reliant on verbal memory, such as history or geography
- supported use of computers, as the auditory requirement is minimal and there is good scope for using visual cue such as attractive graphics and so on

**Please note:** It is almost certainly be necessary for a child to have one-to-one classroom support in order for these recommendations to be implemented.

**Poor attention and concentration** – Many of the above recommendations will apply but the following may also be helpful:

- a quiet and distraction-free classroom environment as far as possible
- small class sizes
- structure the day so that tasks requiring the most attention are scheduled for the time or the day when the child is most attentive, usually in the morning
- give plenty of opportunity for positive feedback
- ensure that you have the child's attention before presenting them with a task
- organisational prompts, such as to pick up worksheets or take certain things to the next lesson
- start with very short periods of sustained focus and gradually increase
- reward periods spent concentrating on work with short period of 'relaxing' with something the child finds easier and more enjoyable, for instance non-verbal tasks



## Further information and support

### Within GOSH

**Developmental Epilepsy Clinic,**  
The Wolfson Neurodisability Service  
Tel: 020 7405 9200 ext. 1144  
Website: [www.gosh.nhs.uk/gosh/clinicalservices/Neurodisability/](http://www.gosh.nhs.uk/gosh/clinicalservices/Neurodisability/)

### Support group

#### FOLKS

**(Friends of Landau Kleffner Syndrome)**

Website: [www.friendsoflks.com](http://www.friendsoflks.com)

Email: [info@friendsoflks.com](mailto:info@friendsoflks.com)

A list of useful contacts can be found in our information sheet *An introduction to Landau Kleffner syndrome: information for families*, available from the Developmental Epilepsy Clinic team or on our website.

## Further reading

Contact a Family – Special Educational Needs factsheet

Available from [www.cafamily.org.uk/educatio.pdf](http://www.cafamily.org.uk/educatio.pdf) – with versions for Scotland and Wales also available from the publications page [www.cafamily.org.uk/publications.html](http://www.cafamily.org.uk/publications.html)

The following books may be available from your local public library. You will probably need the ISBN number if you want to order it from another branch of the library.

- Linda A Hodgdon (1995) Visual strategies for improving communication: practical supports for school & home. Quirkroberts Publishers. ISBN 0 961 678 615
- Susan E Gathercole and Tracy Packiam Alloway (2008) Working memory and learning: a practical guide for teachers. Paul Chapman Publishing. ISBN 1 412 936 136

### Notes

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in collaboration with the Child and Family Information Group

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[www.goshfamilies.nhs.uk](http://www.goshfamilies.nhs.uk)

[www.childrenfirst.nhs.uk](http://www.childrenfirst.nhs.uk)