



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

Abilities and behaviour in Landau Kleffner Syndrome

This information sheet from Great Ormond Street Hospital (GOSH) is part of a series on Landau Kleffner Syndrome (LKS) and focuses on abilities and behaviour. Other topics covered in this series include:

- An introduction to Landau Kleffner syndrome
- Landau Kleffner syndrome: language and communication
- Landau Kleffner syndrome: education

In addition to language, children with LKS often experience difficulties in other areas of development. These areas can impact on a child's ability to learn and interact with the world around them, as well as their psychological well-being and self-esteem. The following section discusses key areas of difficulty in relation to learning, motor skills and behaviour.

Non verbal abilities

Everyone has abilities that help them to understand the world around them, often called non-verbal or performance skills. In children, this includes skills such as visual matching, drawing, design, construction and geometry. In general, these abilities are not severely affected in children with LKS and children can use this area of strength to support their learning, compensate for their language difficulties and maintain 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.

Working memory

Children who are still able to use speech often have difficulties in short term memory and working memory for spoken language. The term 'working memory' refers to the ability to store and manipulate information in your mind over short periods of time. Working memory is important for many aspects of learning including reading and mathematics. Children with working memory difficulties can struggle with skills such as following spoken instructions, doing mental arithmetic and copying from the whiteboard.

A variety of strategies can be used at home and at school to reduce the effects of working memory difficulties. Information and strategies are available in this downloadable pdf guide: www.mrc-cbu.cam.ac.uk/wp-content/uploads/2013/01/WM-classroom-guide.pdf

Executive function

Executive functions are the cognitive processes that allow us to organise our thoughts, feelings and behaviour; and adapt to changing situations. They allow us to plan, start and monitor actions, stop inappropriate actions, and select the information on which to focus. Executive function skills can be affected by LKS and it is not uncommon for difficulties to become more apparent as the child grows older, particularly as they move on to secondary



school. Children with executive function difficulties may struggle with skills such as getting started on tasks, planning what is needed for an activity, completing homework or organising their belongings.

Strategies for supporting children and young people with weaknesses in 'executive skills' are given in the following book: 'Smart but Scattered' by Peg Dawson and Richard Guare [ISBN 1 593 854 455]

Attention

Many children with LKS have poor attention or over-activity. 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. For some children these difficulties are severe, consistently affecting all activities and meeting criteria for a diagnosis of attention deficit hyperactivity disorder.

Attention deficit hyperactivity disorder

Symptoms of attention deficit hyperactivity disorder (ADHD) include inattention, hyperactivity, impulsiveness (not thinking before doing or saying something), little sense of danger, aggression, mood changes and lack of inhibition (failure to control inappropriate behaviour). These behaviours are often due to the disease process, which means that the child probably has little control over these aspects of their behaviour. 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 a child from 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®).

GOSH has produced information on ADHD (including guidance for teachers) which is available online at www.gosh.nhs.uk/medical-information/search-for-medical-conditions/attention-deficit-hyperactivity-disorder/attention-deficit-hyperactivity-disorder-information.

Motor development

Many children with LKS will continue to develop their motor skills as expected, and they may be a relative area of strength. Others may have problems with movements, particularly during the active phase of the condition. Difficulties may include lack of coordination (dyspraxia), shakiness or tremor, unsteadiness, jerky movements, unusual limb postures, weakness or even neglect of one side of the body. When these difficulties are present, they may affect activities such as writing, dressing, or walking and may make it difficult for a child to use gestures or signing. The muscles around the mouth and throat may also be affected, which may cause difficulties with eating, controlling saliva, and speech.

In some cases, the child may experience motor weakness or loss of speech following a clinical seizure. These immediate post-seizure difficulties usually get better after a few hours or, occasionally, days.

If you are concerned about your child's movement abilities, it may be helpful for them to be seen by an Occupational Therapist. If you have particular concerns about difficulties relating to eating and drinking, ask for a referral to a Speech and Language Therapist who specialises in this area. Your local Child Development Centre should be able to help you with both referrals.



Behaviour

At least half of children with LKS show behaviour that is difficult to manage. While some behaviour difficulties may be as a result of the confusion felt by the child at their loss of language, there also appears to be a direct effect of the LKS disease process on behaviour. Furthermore, emotional and behaviour regulation difficulties can be caused by steroids which are a common treatment during the active phase of LKS.

Although behaviour challenges in LKS are often a result of the disease rather than conscious control by the child, the child will still require clear boundaries and consistent behaviour management techniques. It is important that these techniques take into account the language level of the child. If the child has very little understanding of spoken language, adapted visual strategies should be used instead.

It is important to consider underlying causes of behaviour challenges. 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 to manage. It is usually helpful to discuss these matters openly with the child's school, so that appropriate boundaries and responses to the behaviour can be agreed to ensure a consistent approach.

Sleep

As LKS is particularly associated with seizure activity during sleep, it is perhaps not surprising that many children have problems at night. They may have difficulty settling to sleep, have prolonged episodes of wakefulness during the night, have bedwetting or be woken by seizures. Some medications, such as lamotrigine, may also disturb sleep.

Children who have difficulty getting off to sleep may be helped by melatonin. 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, calming bedtime routine, such as a bath, warm drink, reading or looking at a picture book.*
- *a restful bedroom environment, which is dark, quiet and comfortable. Avoid use of screens, such as tablet computers, mobile phones or television, in the hour before bedtime as these can be over-stimulating.*
- *helping a child to sleep in their own bed in their own room, with a baby monitor if you are concerned you will not hear them when asleep.*
- *providing comfort and brief 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 they are asleep.*

Impact on families

The changes in abilities and behaviour often have a great impact on family members, as their child can seem very different in a short space of time. For example, a previously calm and interactive child can become very active and frustrated, leading to behaviour that is difficult to manage. Moreover parents can struggle to communicate with their child and so previous ways of managing difficult behaviour no longer work.

The changes due to LKS can be very difficult for families to come to terms with, and it means that families need to be able to accommodate for and accept their child's current functioning. For children with LKS, their skills and behaviour also change over time and so there are periods in which they show improvements and then sometimes setbacks.

It is important that families seek out support for managing difficult feelings that they are experiencing, and look after themselves both physically and emotionally. This means that they will be in the best position to be able to support their child.



It is also very important to recognise the impact LKS will have on siblings and to access support for them too. For example through Young Carer organisations – details available online at www.carers.org

Financial support may also be available as the child may be entitled to a Disability Living Allowance and the parents may be entitled to a Carer's Allowance. Information is available at www.gov.uk/disability-living-allowance-children/overview.

Further information:

A list of contacts for further information and support can be found in our leaflet *An introduction to Landau Kleffner syndrome: information for families*.

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