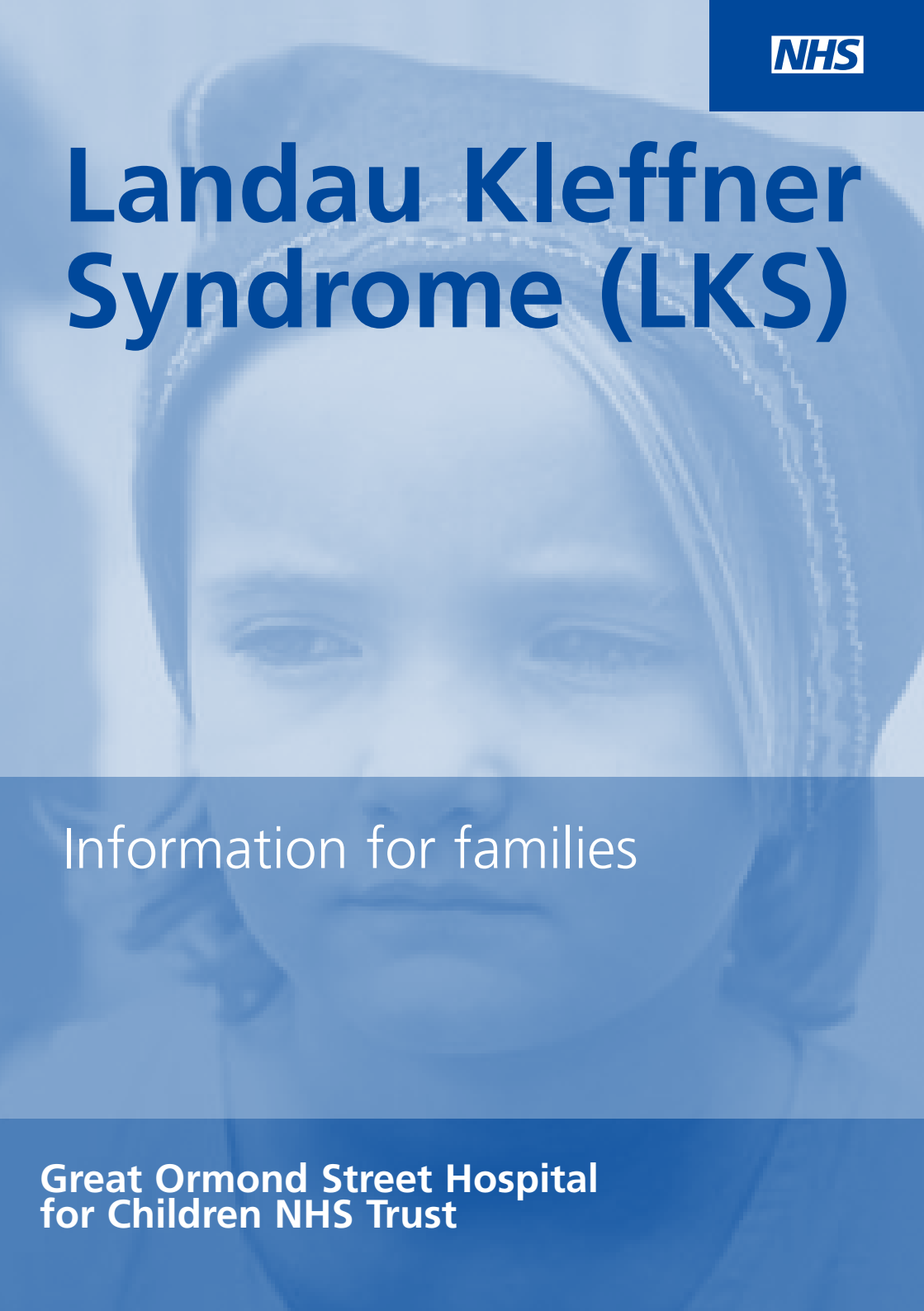


Landau Kleffner Syndrome (LKS)

Information for families

Great Ormond Street Hospital
for Children NHS Trust

Contents

Landau Kleffner Syndrome (LKS)

Landau Kleffner Syndrome (LKS) is a rare form of epilepsy that only affects children, and causes them to lose their understanding of language. The main epileptic activity happens during sleep and is usually not obvious to others. It can be seen on brain wave recordings (EEG, electroencephalography.). There may, however, also be visible seizures at night and/or during the day. LKS may also be referred to by a variety of related terms that describe its effects (see page 49).

As the condition is not well known and has complex effects on language and often also on behaviour, it can take some time before the whole picture is recognised both by parents and professionals and so it can take some time before LKS is diagnosed.

What happens to the child?

In most cases, the child has normal early development, including normal development of speech and language. Onset of the disease is usually between three and nine years and the child experiences deterioration in speech and language ability (see page 16). This loss may be abrupt or gradual over a period of weeks or months and is often initially mistaken for deafness. Many children compensate naturally for the loss of language by using visual cues and gesture, and may initially hide the extent of their difficulty. The deterioration in skills is often called a regression, as the child appears to have returned to an earlier stage in their development.

There are often associated behavioural changes (see page 25) including over-activity, reduced concentration span, irritability, tantrums and difficulties with social interaction (see page 17). The child may also have problems with fine motor co-ordination and movement (for example, dribbling, messy eating, loss of speech clarity, clumsiness and tremor). These difficulties are thought to be a direct result of the disease process, and not simply an emotional reaction by the child to their loss of language.

Most of the children have clinically obvious seizures, and these often start before the initial regression.

The course of the illness is very variable. It isn't usually life threatening, but can greatly affect a child's functioning. Some children may recover spontaneously, while others may recover with the use of anti-epilepsy drugs (AEDs) including corticosteroids, or even brain surgery. Recovery may be complete but more often, children have some degree of persisting difficulties with language, behaviour or cognitive skills. The active phase of the disease often lasts some years until adolescence. During the active phase there may be repeated episodes of regression and recovery, and a child's

understanding and performance may be highly variable even within the same day. The variation can be related to the dose of corticosteroids and attempts to wean them. There is the impression that for many children, the first regression is the most severe, however it isn't unusual for children to recover their skills, only to lose them again in a further regression.

More information regarding treatment is given on pages 9 to 13 and prognosis (or outcome) is discussed further on page 40.

Some children have similar EEG abnormalities as in LKS, but lose skills in all areas (including general intelligence), not specifically in language. This broad group is usually referred to as Electrical Status Epilepticus during sleep (ESES) or continuous spike-and-wave discharges in sleep (CSWS). LKS (in which language is mainly affected) is effectively a specific type of ESES.

We recognise at least two variants of LKS:

- those who had a mild degree of early (developmental) language delay but who showed typical LKS regression later
- those with an abnormality on scan but otherwise a typical history.

The diagnosis of LKS does not include children under the age of two years who regress as part of an autistic spectrum disorder, even if they have seizures or discharges on an EEG. This is because experience has shown that these children fit best within the autistic spectrum of disorders, and do not conform to the pattern of disorder seen in LKS.

Why does it happen?

Very little is known about the causes of LKS. The condition is twice as common in boys and only very occasionally runs in families. It may be that there is a genetically determined vulnerability, which becomes apparent in response to an environmental trigger, for example, infection, but there is as yet no scientific evidence for this.

How does it happen?

All children with LKS can be shown to have seizure activity during the active phase, that usually affects both sides of the brain (although one side may seem more affected), and is often concentrated in areas known to be important for language (centro-temporal region). Some of this activity results in actual seizures but much of it does not, that is, it is 'sub-clinical'. EEG recordings show that there is a particularly high rate of sub-clinical epileptiform activity in sleep, which often amounts to nearly continuous spike-and-wave (CSWS) discharges (Electrical Status Epilepticus during Sleep or ESES) during the active phase of the disease.

It is thought that regression and impairments are related to these epileptiform discharges during sleep, and that these electrical seizures 'short-circuit' the normal wiring so certain functions of the brain are prevented. This seizure activity, which is often-widespread, prevents the child from using his or her brain normally so they regress in abilities. Initially, the brain is not 'damaged' in the conventional sense, but rather caught up in an 'electrical storm' that blocks certain brain functions (especially language, attention, social functioning). Stopping seizure activity may restore these functions.

LKS mainly affects a child's language abilities, and this is probably related to the common location of recorded discharges over the key language areas (centro-temporal region). It was initially thought to be specific to language, but certainly current experience is that other higher

functions are also commonly affected, including attention, social interaction, behaviour and motor control. Non-verbal cognitive skills are usually relatively spared, although not always, and it is not unusual to have specific or more general learning difficulties.

Unlike physical injury where brain 'plasticity' allows other areas of the brain to take up important functions, in LKS, the brain's capacity and reserves appear to be limited by the electrical activity. Consequently, relocation of skills (such as language) to other brain areas is not generally possible.

The active phase of the disease relates to the period of sub-clinical seizure activity and appears to be time-limited, starting after the age of three, and 'burning out' by early adolescence. The visible, clinical seizures are generally short and do not show a close relationship with the effects on language and other areas of development.

This 'seizure mechanism' that produces the deficits, makes LKS (and related epilepsies) quite unlike more common developmental disabilities, which are usually present from birth with static deficits affecting all aspects of learning evenly. LKS is also quite different from traumatic brain injury where there is actual damage to brain substance, usually visible on a brain scan, with predictable loss of abilities related to the damaged areas. The brain-injured child usually makes steady progress once they have recovered from the immediate injury, and in some cases, the uninjured brain areas may take over the lost skills.

LKS and related severe seizure disorders are unique in causing extreme fluctuation because of the variable nature of the electrical activity. A child's understanding and abilities may change dramatically (for better or worse) over short periods of time, and for some children, there may be obvious variation even within a day. This poses a major challenge for those supporting the child particularly in the classroom (see page 33).

How is it diagnosed?

LKS is a clinical diagnosis, which means it is made on the basis of the child's history and assessment. The core features are a history of normal early development followed by loss of language skills, often in association with mild observed seizures and behavioural changes. There is no specific test, although EEG recordings can be very helpful, especially in the active phase of the disease. MRI scans are usually normal.

The condition is rare and may not be thought of initially. It is common for children to be investigated for deafness, autism, selective mutism, verbal dyspraxia or behavioural problems before the diagnosis is made.

Your child will have an initial medical assessment, including examination. The physical examination is usually normal apart from occasional mild co-ordination or other movement problems. The doctor may request tests to check for various alternative diagnoses. The tests are typically normal, apart from the EEG.

There will also be assessments of your child's development across different areas of learning, particularly language. It is important to record your child's current skills as a baseline, which can be used to gauge the effect of the disease and any medical treatment or therapy, in the future. This assessment will also allow the therapist to identify appropriate intervention(s) for your child (for example, speech and language therapy). Your child should then have regular assessments to monitor changes in skill profile. This information will be important for making decisions about medical, educational, behavioural and therapeutic management.

It is important that your child is assessed at an early stage by a multidisciplinary team including medical, speech and language and clinical psychology services. This enables your child's full profile to be assessed and considered in the management programme, and a co-ordinated approach to be adopted by all people working with you and your child.

What tests may be done?

MRI (Magnetic Resonance Imaging) brain scan

This produces a very detailed image of the brain. Your child has to lie inside a machine, which is like a small tunnel, and can be noisy. The machine uses a big magnet and radio waves to take a picture of the brain, a bit at a time. Then a computer creates the picture. It does not involve X-rays. The scan takes quite a long time (up to 40 minutes) and so many children will need either sedation or a general anaesthetic to help them to lie still. For most children with LKS, the scan appears normal.

CT (Computer Tomography) brain scan

This also produces a picture of the brain, but it is less detailed than MRI. It uses X-rays, and is much quicker to perform but is not the preferred imaging mode for epilepsy.

EEG (Electroencephalogram) brain wave record

The EEG is a special test that records the electrical activity from the brain. It is used particularly to look for clues about fits. Your child has wires stuck onto his/her head with special glue, which record electricity coming from the brain (it is simply recording the brain's normal activity). During the recording, your child will be asked to open and shut his or her eyes, and at one point to breath deeply (or blow a windmill). He or she will also be asked to look at a flashing light. If possible the recording will include a period of sleep, which is particularly important to monitor with LKS. In some children these activities may increase or reveal abnormalities, which can then help to guide the medical treatment.

During the active phase of LKS, EEG recordings will usually show abnormal discharges on both sides of the brain over the centro-temporal regions, and these discharges often become

continuous in sleep. Therefore a sleep record is usually required when assessing a child with LKS, and often this will be achieved by a period of video-telemetry (typically overnight).

Video-telemetry means using a closed circuit video camera, which is linked to an EEG machine. The camera records what is happening to the patient at the same time as the EEG records the brainwaves, and the screen displays the patient and the EEG trace simultaneously.

If surgery is being considered, the following specialised tests may be used:

Methohexital suppression test

In this test, your child is monitored using EEG. A light anaesthetic is given and a short-acting barbiturate drug (methohexitol) is given to put your child deeply asleep to the point where the EEG recording of brain activity becomes a flat line. The drug is then allowed to wear off, and the EEG begins to show electrical discharges again. The first place where this activity returns is thought to be related to the source of the seizure activity. This information is helpful in planning surgery.

Magnetoencephalography (MEG)

This detects tiny magnetic fields that are part of seizure activity, and is thought to localise the seizure source very accurately. It is particularly helpful if the seizure source appears to be located in one of the folds of the brain's surface (commonly the sylvian fissure in LKS) as it gives a three dimensional localisation which is superior to EEG information. However, the equipment is expensive, bulky and not currently available for children in the UK although there are plans to address this. If this test were needed, your child would currently need to travel to Helsinki.

Single Photon Emission Computed Tomography (SPECT):

A radio-labelled tracer is injected into a vein (often using a 'plastic drip' that has been inserted into the back of the hand), and the brain's uptake of the tracer is measured, with the seizure source showing reduced uptake to the rest of the brain.

What treatment is available?

Management can be divided into two categories:

- treating the seizures and seizure activity, thereby trying to change the disease process and reduce its effect on your child
- providing functional support to optimise recovery.

The first category is described below. Strategies from the second category are described within the relevant sections on pages 17 to 39.

Medical treatment

As described earlier, there are two aspects to the seizures in LKS

- the observable 'clinical' seizures which do NOT appear to correlate with severity of the developmental impairment
- the electrical seizure activity that occurs in sleep and is thought to cause the regression

Antiepileptic drugs (AEDs) or anticonvulsants are drugs that are used to stop seizures. They are usually very effective for the

visible seizures but their effect on the sub-clinical seizure activity, which is characteristic of LKS and typically occurs in sleep, is often disappointing. Some children may respond to conventional AEDs, and it is well recognised that high dose benzodiazepines (for example, clobazam taken usually at night) can be particularly effective. Sodium valproate is also commonly used and occasionally other AEDs appear to be effective.

Corticosteroid drugs can be dramatically effective in stopping seizures and reversing a child's losses. They are either used in short high-dose courses or in prolonged weekly (pulsed) courses with careful monitoring of side effects. Some children recover well with a single short course (steroid-responsive), others make good recovery but lose skills when steroids are stopped (these children are steroid-dependent and may respond to longer-term weekly steroids). Others have only partial or no response. The most complete steroid responses appear to be seen in children whose regression is largely limited to inability to understand speech (that is, pure auditory agnosia) and who do not have additional impairments in behaviour, social communication, cognition etc.

As with all treatments, it is important to consider the benefits and risks involved, and to be clear about the aims. LKS is notoriously difficult to treat, so it is important to have evidence of a treatment's effectiveness, before subjecting a child to prolonged medication. Specialist assessment by a speech and language therapist, before and after starting treatment is very helpful in documenting changes in skills and judging effectiveness. Baseline assessment of cognitive skills is also very useful in determining the overall learning profile and identifying strengths and weaknesses.

All drugs have side effects and it is necessary to monitor for these (for example, steroids – sugar in urine, blood pressure). Steroids, in particular, are powerful drugs that when given in high dose on a daily basis can affect a child's growth, bone strength, ability to fight

infection and lead to diabetes, high blood pressure and even stomach ulcers. This is why daily steroids are usually restricted to a short period (such as six to twelve weeks). Weekly pulsed steroids appear to allow the medical benefit, without the same side effects. However, chickenpox is a serious illness if a child catches it whilst on any form of steroids. It is important to discuss this with your doctor if your child has not had this infection and we recommend immunisation before treatment.

Many parents worry about the effects that drugs have on learning but this is rarely a problem. It is the subclinical seizure activity that has the major impact on learning, and generally, drugs that control this activity enable learning to take place without ongoing interference from seizure activity.

Some parents also express concerns about the possible behavioural effects of drugs (for example, drowsiness, overactivity, changed appetite, insomnia, bedwetting). This can be a problem, and children with LKS appear particularly vulnerable to some side effects such as irritability with sodium valproate, or sleeping problems with lamotrigine. It is often hard to disentangle these from the behavioural difficulties commonly seen in children with LKS. For example, it is not unusual for parents to describe increased aggression and hyperactivity associated with the early phase of steroids – although equally, many parents report dramatic improvement in their child's behaviour on steroids as the disease comes under control. Specific concerns should be discussed with the local team managing the child.

It is important to realise that all drugs have two names, as this can be confusing. There is the generic name (main chemical the medicine is made from) and the trade name (used by the company which produces the drug). For example, sodium valproate is the generic name and Epilim® is the trade name. It can be difficult to persuade children to take medication and different formulations such as syrup or sprinkles may be helpful.

There are also occasional anecdotal reports of benefits from other treatments such as immunoglobulins or a ketogenic diet.

Surgical treatment

Brain surgery is occasionally used in LKS to limit the effect of the seizures.

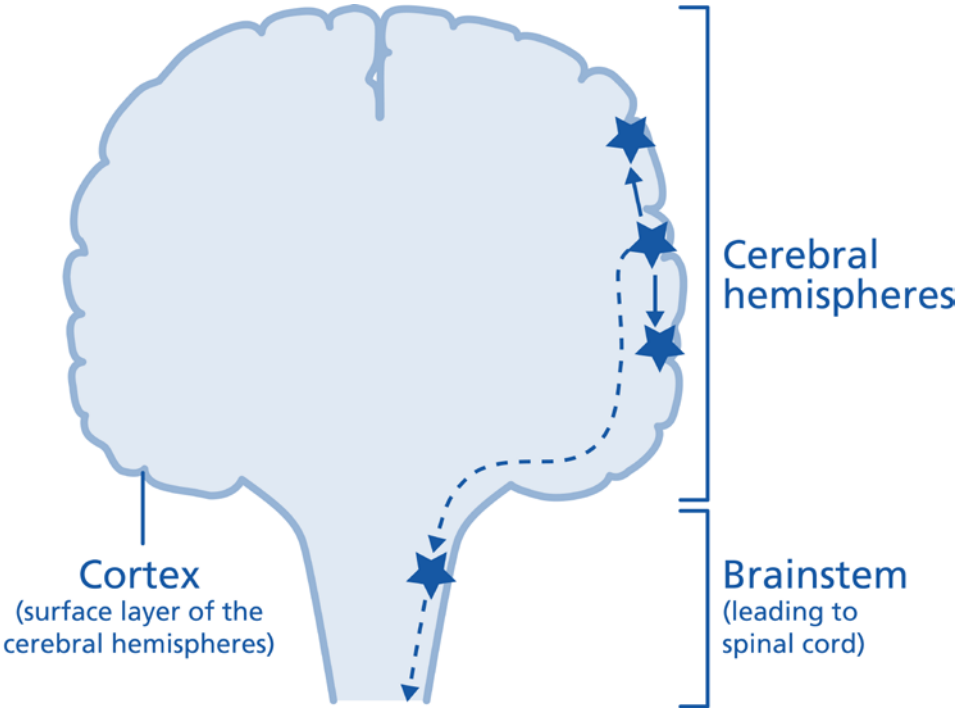
The surface of the brain (cortex) is organised into specific areas that deal with special functions such as movement or language. Brain cells (neurones) in this surface layer, have important fibres that pass through the brain substance to carry messages to control the rest of the body. These brain cells also have small fibres that branch out and connect them to the other brain cells in the surface layer. In LKS, one area of the brain's surface develops electrical discharges or seizures. This area then spreads the seizures to other areas of the surface, through its network of small fibres, and thereby becomes 'dominant' and 'drives' the rest of the surface or cortex into discharges that 'tie-up' the brain cells and prevents them from carrying out their specialised function, such as language.

Brain surgery for LKS aims to prevent spread of seizures through this surface network by making tiny cuts over the surface where the seizures originate, preventing the discharges travelling sideways to other surface areas, whilst preserving the long fibres that carry the specialist messages to the rest of the body. This surgery is called 'multiple subpial transection' and requires specialised assessment to identify the 'dominant driving' area of the brain surface to be targeted.

Brain surgery may be used for children who have active disease with poor recovery of skills and EEG evidence of continuous seizures in sleep, or for those who require unacceptably high doses of steroids to maintain their recovery. It does not aim to cure the child, but to limit any further loss of skills and allow some recovery.

Less than half of the children who are assessed for surgery, are found to be suitable on investigation.

Brain surgery inevitably has some risks. However, the experience in reputable centres is that more than half of the children experience significant improvement, not simply in language, but often most markedly in behaviour, particularly autistic features. Brain surgery however, is not curative and the children will have some remaining impairments, although experience to date is that no children have been made worse by the procedure.



Surgery (MST) aims to cut the surface fibres (solid arrows) and hence prevent the spread of seizures to other surface areas, whilst still preserving the long fibres (dashed arrows) which take messages to the rest of the body.

The Clinical Care Pathway

The NICE (National Institute for Clinical Excellence) Epilepsy guidelines (Oct 2004) recommend:

- early referral to a paediatrician with special responsibility for epilepsy (within 2 weeks of first seizure)
- development of a comprehensive care plan
- regular review
- referral to tertiary services if there is diagnostic uncertainty or treatment failure

The services should be child-centred, and the review should provide access to written and visual information about their condition, counselling services, voluntary organisations, epilepsy nurse specialists and integration with other community and multi-agency services involved in children's education, welfare and well being. This integration may commonly be mediated by the epilepsy nurse specialists.

Many of these recommendations are very appropriate in LKS, once the diagnosis has been made. However, there are difficulties as these guidelines refer to the case of clinically apparent seizures, which do not always occur in LKS (and in any case, are not the main problem). Furthermore, there is often a significant time delay before the diagnosis of LKS is made, so care pathway recommendations must be slightly modified, as below.

1. **Early referral to a paediatrician** should be triggered either by a seizure OR loss of language abilities without overt seizures. In LKS, children demonstrate loss of previously acquired language abilities in association with subclinical seizure activity, although this may initially be mistaken for other conditions (e.g. mutism, deafness, behavioural problems).
2. A paediatrician should conduct an initial assessment and investigation. Ideally, this should be a **multidisciplinary**

assessment by the local team, including speech and language assessment, and assessment of cognitive abilities / developmental level. A paediatric neurologist would usually be involved in further assessment of such a regression, and would arrange specialist investigations (e.g. sleep EEG or video telemetry) and a multidisciplinary assessment as necessary.

3. After diagnosis of LKS, a **paediatric neurologist would generally oversee the child's medical management** and liaise with the local paediatrician, who would be responsible for coordinating therapy and support for the child and their family.
4. **Regular review** during the active phase of the disease would involve close liaison between paediatric neurologist and paediatrician, and the facility for language and cognitive assessments (particularly to monitor response to changes in medication). There should be access to advice on appropriate educational placement, and behaviour management if necessary (child psychiatry / psychology).
5. **The child may be referred on to a specialist paediatric epilepsy centre** (such as the Developmental Epilepsy Clinic at GOSH) if there is:
    a. poor response to treatment
    b. further loss of skills or 'plateauing' in development
    c. complex or severe behaviour problems
    d. the possibility of epilepsy surgery

The Effects of LKS and Therapeutic Stratgies

Language and communication skills

LKS encompasses a broad spectrum of children with varying degrees of language difficulties. During the course of the disorder, it is not uncommon for language skills to fluctuate especially when the EEG abnormality is not controlled (see page 4).

Language problems are usually first characterised by difficulties in understanding spoken language. As mentioned earlier, hearing loss may initially be suspected but formal testing (pure tone audiometry) invariably confirms a normal hearing system. The difficulty lies in the interpretation of the sounds. The difficulties with comprehension vary from problems understanding complex and longer instructions to complete inability to understand spoken language, including loss of understanding of previously known, simple vocabulary. In some children, the problems may become so severe that even environmental sounds (such as a dog barking, a telephone ringing, traffic noise) lose meaning for the child.

Difficulties with spoken or expressive language typically follow and show themselves in many different ways. For those who are still able to speak, sentences may be simplified and reduced in length. Some children experience problems retrieving known words from their memory (a “tip of the tongue” experience). Their spoken language may consequently contain many pauses as they try to find the word or they may substitute alternative words (for example, writing stick for pencil). Some children slot the incorrect sounds into words so that the word produced resembles the target but is not a real word (for example, gilat for giraffe). Speech may also be affected with changes to intonation or voice quality. Some children sound slurred or speak in a jerky, hesitant manner. The spoken

language difficulty can become so severe that the child does not have any speech at all. In such cases, the child may resort to using gesture or mime to communicate. Reading may or may not be preserved in children who had previously acquired this skill. More than half of children with LKS also have difficulty using gesture.

In some children, social functioning may also be affected, with problems resembling those of children with autistic spectrum disorders (ASD). This may or may not amount to an additional diagnosis of autism or ASD, which is made on the basis of a pattern of difficulties observed in the areas of social interaction, social communication and imagination. Individuals vary in how it affects them but features can include loss of desire to interact, self-directedness and problems with eye contact and facial expression. In addition, children may have difficulty using natural gestures or signs to communicate or they may use their communication skills only when highly motivated to get something (that is, needs driven, such as wanting a drink when they are thirsty) rather than just for social reasons (for example, to draw attention to an object of interest or share pleasure). At the milder end of the spectrum, problems may be noted with conversational skills and more subtle aspects of interaction (for example, understanding and producing sophisticated facial expressions such as guilt, embarrassment). This group of symptoms is considered in more detail on pages 22 to 23.

Language therapy and educational setting

Since language for the purposes of learning and communicating is closely related to context, the environment is a critical factor in the successful management of language problems associated with LKS. Some experts have pointed out that creating a situation where the child feels at ease is a key aspect to the rehabilitation programme as it sets the scene for enthusiastic learning. Others

have advised that language therapy should be integrated into classroom management. Language therapy and educational management should therefore always be considered in relation to the other.

Educational placements will be covered in more detail in the 'school' section (pages 33 – 39).

Speech and language therapy

Speech and language therapy is an important part of the management of children with LKS. It should be delivered as part of a global approach, which also includes medical intervention, educational management, behaviour management (if needed) and pastoral care. Knowledge of the child's general cognitive skills is essential to ensure an appropriately tailored programme, which gives consideration to all of the child's abilities. It is also important for the speech and language therapist to work in conjunction with the medical team and to be aware of changes in medical treatment as language assessments can help to determine the effectiveness of these interventions.

It has been suggested that speech and language therapy should be provided as soon as possible after the onset of the disorder. All intervention must be tailored to the individual needs of the child and must include a high degree of flexibility and responsiveness. Many children are very variable in their performance, and the disorder is also very changeable, so it is inevitable that any programme will need frequent review and adaptation. Input during the early stages or following medical intervention should be intensive and have high priority to maximise the child's potential for progress. Speech and language therapy is likely to be necessary (for the majority of children) on a long-term basis. LKS is a rare syndrome and specialist advice should be sought as required to help determine the most appropriate form of intervention.

It is appropriate for restoration and development of spoken language comprehension and expression to be a first goal for the intervention programme. This may, however, need to be adapted depending on the responsiveness and progress of the child over time. Consequently, a broad based functional approach that builds on residual skills while maximising the child's strengths is recommended. Such a pragmatic method enables the development of a range of communication skills and therapy goals can be adapted depending on the child's progress over time. For children with severe language problems, the focus may be on providing an alternative means of communication (for example, symbols or signing) to ensure that the child can still communicate their needs and interests. For those with mild to moderate language problems, the focus would be on developing areas of weakness to facilitate the child's ability to communicate more successfully through spoken language.

Visual cues and alternative communication

Visual cues are an important support as the brain still processes visual information relatively normally and this can therefore be used to compensate for problems with processing auditory information. This enables the child to communicate despite their difficulties with spoken language and this can reduce the frustration and behavioural problems that so frequently arise. Visual cues take a number of different forms including signs, pictures, symbols and written language etc. Some children with LKS have particular difficulties (for example, with gesture, interpreting visual cues including facial expression or lip-reading) that can make it very difficult for them to use some of the alternative communication methods.

Children in the early stages of the disease or those who have not regained sufficient spoken language to enable them to use this functionally, may benefit from signing which has the advantages

of being quick, portable and not dependent on having specific pictures or symbols to hand. Experience with children who sign shows that this will not prevent them from developing spoken language if they are capable of this and indeed, there has been some suggestion that it may even help to promote it. Some children benefit from systems such as Makaton, which provide basic levels of signing. Others progress beyond this to a more sophisticated system such as British Sign Language which enables them to express themselves using complex language. Signing is not successful in all children and should therefore be monitored carefully to determine its usefulness. Nevertheless, gauging the success of signing is dependent on providing the child with adequate opportunity to learn the signs and it is important that these are used consistently across the whole day and in all contexts (for example, home and school).

For children with more significant learning difficulties or autistic spectrum type problems, the use of concrete visual cues such as objects, pictures or symbols can be very helpful. Visual cues provide the child with more time to process the information compared to spoken language or sign language and tend to look like the object they are representing thereby providing the child with more concrete clues.

Visual cues can also be used in a way that helps to make the two-way nature of the communication process more explicit (for example, handing a picture of the desired object to another person) which is important for children who do not readily understand this process. The Picture Exchange Communication System (PECS) is an example of a programme, which aims to develop the underlying understanding of the communication process. Children are taught explicitly about the 'give and take' nature of communication through explicit demonstration of this process by actually handing over a picture or symbol as they make their request. PECS also encourages children to initiate communication rather than wait for others to approach them. It is important to choose highly motivating material.

Cued articulation involves the use of simple hand signs to show the position of the tongue for consonant sounds in children who have articulation problems.

Written instructions can be useful as a means of supporting or supplementing spoken instructions in children who can read. Those who are being taught to read may benefit from the additional use of colour to reinforce the different categories (nouns, verbs etc) as described by Lea. Vance also described the process of 'graphic conversation' to develop reading skills through the use of speech balloons to record a child's story. See the Reading list on page 48 for further details.

It has been suggested that a visual rather than a phonological approach to teaching reading may be best. This would mean teaching the child the whole word at once (usually written under the visual symbol or picture, or even stuck to the real object) and allowing them to recognise the overall pattern of the written word, rather than sounding out the individual letters and then trying to blend them and pronounce the word. Once a child has reading skills, this in turn can be used to improve auditory analysis.

Auditory training

Many children with LKS have short-term auditory memory problems as well as problems processing individual sounds within words (an important skill for acquiring literacy). Strategies such as repetition of spoken instructions, reducing speech rate and background noise or distraction are particularly important for classroom management. Specific auditory training has been used alongside more traditional therapy to develop the skills, which underpin language development. Some have recommended the use of FM amplification systems with a low gain output in the classroom as a means of helping the child to focus on the class teacher's voice. This does however make it difficult for the child to engage in classroom discussion with peers, and may be difficult for the child to tolerate.

Social interaction and communication

As mentioned earlier, children may experience difficulty with aspects of social interaction and communication. This will impair their ability to relate to peers and form or maintain friendships. They may continue to show pleasure in certain activities but fail to share this pleasure with others through language or other communication modes (for example, eye contact, facial expression). They may make inappropriate remarks or behave in socially unacceptable ways with little awareness of the social implications of these behaviours. For children who have very little language, there may be failure to compensate for this problem by gesturing or miming in order to get their message across. Some children who are able to produce language may have difficulty using their language socially or engaging in a two-way conversation. They may echo language around them, or reproduce set learnt phrases in an inappropriate way. In addition, these children may have problems with abstract thought or generalising from experience. They may have difficulty with imaginative play and some children show obsessional and repetitive behaviour. They may also find unstructured situations, such as the playground, and periods of change or transition very difficult, preferring to stick to familiar routines.

Although these problems are acquired usually around the time of the child's illness rather than being developmental (that is, present since infancy), they share many similarities to children with autistic spectrum disorders. For some children an additional diagnosis of autism / autistic spectrum disorder may be appropriate whilst in other cases, their behavioural features will not amount to a full diagnosis, but remediation strategies relevant to this population may nevertheless be recommended.

A general emphasis on the use of structure including daily schedules as described in the TEACCH approach (Treatment and Education of Autistic and Related

Communication handicapped Children) can be useful in terms of its ability to convey meaning, predictability and order to the child. The use of visual cues (see page 19) can be very useful.

Social skills training may be useful for children who are experiencing problems with social interaction and communication. The evidence (based on children with autism) suggests that there is often a perceived benefit by the child and parents although these skills can be very difficult to teach and transfer to everyday situations. In addition to formal training, many children benefit from support aimed at providing the child with skills to use in social settings (for example, teaching games which can then be re-enacted in the playground) as well as practical help for specific situations as they arise.

There are also some children who do not have autistic spectrum disorder, but who respond negatively and avoid social situations as an understandable reaction to their loss of language.

It is important to recognise these difficulties as they have significant implications for classroom learning, behaviour and the development of social relationships.

Other cognitive abilities

Non-verbal skills

As described above, LKS causes a significant impairment of language skills, usually in terms of both understanding and speaking. Although verbal abilities are probably our most obvious set of skills, each individual also possesses a range of other 'cognitive' abilities contributing to their intelligence, often referred to as non-verbal or 'performance' skills. As the name suggests, these underlie our non-verbal understanding of the world and in children they include skills such as visual matching, drawing, design and construction, geometry, and mathematical problem solving. These non-verbal abilities may be assessed using a variety of different psychometric tests or intelligence tests. Depending on the child's age these may include tasks such as inset puzzles or jigsaw puzzles, drawing and copying, and 'block design' (constructing a geometric

pattern from coloured blocks). Accurate assessment of these skills can be very difficult, however, if the child's motor skills and/or attention and concentration have been affected.

As a general rule, non-verbal skills are relatively spared by LKS, that is, there is often some impairment, but usually this is less severe than the language deficits (and sometimes there is no measurable impairment at all). This has important implications for the child's education (see school section on pages 33 to 39), as it is important to continue to use these preserved visuo-spatial skills in order to optimise their development long-term, and also to boost self-esteem at a time when many of the child's abilities have been taken away from them. A more severe impairment of non-verbal abilities is sometimes seen, however; that is, equivalent severity to the language impairment, such that there is an even or 'global' pattern of delay in the child's development. The clinical impression is that this picture predominantly affects children in whom there has been an early onset of LKS. Where there has been some significant impairment of non-verbal abilities associated with LKS regression, they are often the first to recover once the child starts to make gains again.

Some specific strategies for supporting children with this pattern of difficulties are set out in the Useful Teaching Approaches section on pages 36 - 38.

Memory and attention

LKS may result in the child having specific difficulties with memory and attention, particularly related to verbal material. If there is a moderately severe degree of general cognitive or language impairment then any such specific deficits may not be measurable. However, for children whose cognitive impairment is not severe (or has recovered significantly) specific memory problems

may become apparent. These specific difficulties are a direct consequence of the abnormal brain functioning that occurs in LKS, particularly affecting the fronto-temporal regions of the brain, which are closely involved in memory processes.

If these difficulties are suspected, a full neuropsychological assessment should be carried out by a clinical psychologist to determine the pattern and severity of the problem. In terms of verbal memory (where problems are most often expected), care should be taken to try and differentiate between problems that stem from the child's difficulty attending to and/or processing incoming information (that is, related to a primary auditory processing problem) and any additional difficulties related to storing this information.

Depending on the pattern of difficulties found, a variety of strategies can be employed at home and at school to minimise the consequences. These include using simple visual mnemonics (memory prompts), timetables, checklists of what to take to school, etc. Some further suggestions are given in the Useful Teaching Approaches section on pages 37 - 38.

Behaviour

It is estimated that at least half of children with LKS experience neuropsychological and behavioural difficulties as a result of the condition. A wide range of specific difficulties has been observed, with the most common categories described below.

Attention deficits, hyperactivity and aggression

Some features of poor attention or concentration, or over-activity affect many children with LKS at some point, and these may be associated with irritability and aggression (that is often towards particular family members) in some cases.

In the most severely affected, these features may be consistent with Attention Deficit Hyperactivity Disorder (ADHD) and the child's ability to engage meaningfully with their environment is markedly compromised. However, in many cases the characteristics are much milder and may only be noticeable to close family members or teachers (the child is a bit more 'bouncy' than usual, has become slightly impulsive, or has difficulty sustaining concentration throughout a whole lesson). In others, the features are marked but episodic, for example, a couple of hours of overactive behaviour in the evening, or may be more pronounced in particular environments, for example, large gatherings where there is a high level of noise and stimulation. The most common features reported are: inattention, hyperactivity, impulsiveness (that is, not thinking before doing or saying something), no sense of danger, verbal and/or physical aggression, mood changes, and disinhibition (failure to inhibit inappropriate behaviour, for example, making rude comments to unfamiliar adults or pulling their trousers down in public).

It is often assumed that these behaviours are purely a response to the frustration felt by the child to the loss of language. Although most children with LKS do experience episodes of extreme frustration and confusion as a result of the condition, there is little evidence to suggest that this is the primary cause of ADHD type behaviours. For example, attention difficulties can present before there is any apparent language deficit. In addition, recovery of most areas of dysfunction, including behaviour, can occur even when significant language difficulty persists. It is therefore thought to be a direct result of the condition (see below).

However, the social and emotional impact of a sudden loss of abilities should not be under-estimated and this factor will almost certainly contribute to behaviour patterns.

Most often, ADHD-type problems will show some improvement associated with improvement in control of the underlying seizure activity during sleep, and with recovery from regression (and conversely, deterioration in behaviour is found to be related to the disease worsening). In some cases, the behaviours will resolve completely and dramatically when the disease is effectively treated. In other instances where hyperactivity is very severe or persistent, it may respond to treatment with medication that specifically targets this group of disorders (for example, methylphenidate or atomoxetine). It is important to treat these ADHD-like difficulties in their own right, as they may prevent the 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.

It is thought that these behaviours primarily result from interference with the brain's normal functions, caused by the abnormal electrical activity that is associated with LKS (whether or not there are frequent overt seizures). This means that the child probably has very little control over these aspects of their behaviour.

However, there is a further acquired element that can also influence the occurrence of challenging behaviours. First, in children with a very long-standing disorder, poorly regulated behaviour may in part reflect the fact that one of the most important channels for teaching/learning such behavioural control (that is, oral communication) is not available. Second, through simple association children may 'learn' that some of these behaviours produce a desirable outcome, for example, if they have a tantrum and throw things around when the TV is turned off, then someone turns it on again. This means that the behaviour will then 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. Studies have shown that behaviour management techniques remain successful in helping this group of children, despite the fact that the behaviours have a significant organic component (that is, are due to the disease process, not simply a secondary psychological reaction to it).

Useful approaches include:

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

Judging whether a child has control over their behaviour or not, can be very difficult, and the advice and input of a local clinical psychologist (often from the Child and Adolescent Mental Health Service or 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 response.

In children with milder difficulties involving more 'cognitive' inattention and impulsivity these strategies may help:

- playing games that require attention and memory to encourage these skills (there are many examples available, for example from Early Learning Centre®) – but particular attention should be paid to the appropriate level of difficulty so the child has the experience of achievement, not failure.

- the parent counting to ten before responding to a situation that is upsetting
- discussing basic rules to help with impulsivity – "Stop & Think"
- creating simple visual mnemonics (memory prompts) to help remember important verbal information.

Sleep disorders

Many children with LKS are particularly active in the evenings and parents report that they cannot settle to sleep until late. In other cases they go off to sleep readily in the evening but then have prolonged episodes of wakefulness during the night, or wake in the early hours and cannot go back to sleep. LKS is particularly associated with seizure activity during sleep so it is perhaps not surprising that so many children have problems at night. Indeed, many parents report that their child is woken by the seizures themselves during the night. Also some drugs (for example, lamotrigine) may disturb sleep.

Children who have difficulty getting off to sleep may be helped by melatonin (it is also used to treat jet-lag in the adult population). It is harder to treat night time waking. It may not be possible to 'cure' the underlying medical reason for these sleep difficulties, but the situation can usually be improved by consistent application of standard behavioural management strategies. These may include:

- a regular, quiet bed-time routine (bath, warm drink, being read a story or shown a picture book)
- removing TVs and videos from the child's bedroom
- sleeping in their own bed in their own room (with a baby-monitor if you are concerned that you will not hear them when asleep)
- comfort and reassure when your child wakes at night but don't overdo it (that is, resist switching on all the lights/ giving food / turning on a video/ staying with them until they fall asleep).

Other behaviours

Some parents report that their child seems very tearful and depressed, and this should be carefully monitored. Understandably, many children with LKS will require more reassurance than usual and may seek physical comforting or become anxious in social situations. Others may become more controlling of their environment.

A small proportion of children become passive and apathetic in their manner. In our experience, this is most commonly associated with a marked global regression and early onset (before two-and-a-half 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.

Motor difficulties

Motor problems are very common, occurring in around two thirds of children with LKS. They often relate to the disease activity (that is, correspond to periods of regression or fluctuation). They may include dyspraxia or inco-ordination, tremor, unsteadiness, jerky movements, unusual limb postures, weakness or even neglect of one side. They may affect activities such as writing, dressing, walking and may make it difficult to use gesture and signing. The muscles around the mouth and throat are commonly involved and will cause difficulties with feeding, controlling saliva and speech.

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

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. But for most of us and for the world around 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 subtly transformed so people use a language you can't understand or speak yourself. You might try to guess what is happening from clues around you, but it will be very tiring and unrewarding.

Because 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.

Children who lose the ability to understand environmental noise, will need special support and supervision. Certain situations will be more dangerous for them for example, as 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 who 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 (for example, tannoy announcements) can be very distressing for the child.

The family provides the main care for the child. Parents are usually with their child most frequently, and are the best source of information about the child throughout the illness. They will often detect change in the child's condition, before it is formally apparent. They accept and nurture the child, provide structure and sense to their world, and will be the main communication partners. They should be actively involved in decisions, and given appropriate information and support, including opportunities to learn special skills (for example, signing, PECS) that can be used at home.

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

Therapists (speech therapists, psychologists, autism advisory service etc) are skilled at establishing a child's strengths and weaknesses, and at identifying the best approaches to support the child. They will work closely with class teachers and assistants and many of their recommendations will be implemented through class work. Regular reviews are important to judge the success of any schemes and to monitor the child for rapid changes in ability. Rapid gains may merit intensive therapy to optimise the recovery phase. Rapid losses will mean that the child needs more support, perhaps even new ways of communicating, and any deterioration should be brought to medical attention.

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 make sense of their world, as well as providing a stable social structure. Given the complex and unusual nature of 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.

Educational challenges

Whatever form of school placement is chosen, a child with LKS continues to pose many challenges, which the school must adapt to, most notably:

1. Their condition can change rapidly over time, that is, 'fluctuate', making progress at school erratic, and support needs to be responsive to this. Regular monitoring and updating of therapeutic and educational plans is necessary
2. When the child's disease is active, performance can vary even within a day, making them susceptible to fatigue and difficulties with concentration. Teachers/LSAs must be made aware of this and careful timetabling of lessons may help to minimise the impact
3. Despite having significant language difficulties, many children with LKS retain average or above average abilities in the non-verbal domain. However, because standard classroom presentation (instruction and so on) is almost invariably verbal, this means that a special teaching approach must be devised (see below). 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

4. Other cognitive effects of LKS such as slow processing and impaired verbal memory make it even harder for LKS children to understand what is required of them. For example, children with LKS may understand language in a quiet one to one situation, but in a noisy classroom the listening environment is very complex and the child may well 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 quite unable to remember a sequence of verbal instructions or a story – which would cause enormous difficulty in class and also with playmates. However, the severity of this difficulty may be masked by the abilities that are preserved and by clever use of well-learned social behaviours (children usually want to cover up what they can't do) and this may be misconstrued as 'naughtiness'. Useful strategies for tackling memory/processing problems are described below
5. 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, aggressive outbursts. (A more detailed description, including suggested coping strategies, is set out in the 'Behaviour' section on page 25 - 30).

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 need to be produced. The statementing process is carried out by your local education authority and may take several months, involving assessments by local educational psychologists and speech and 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 (for example, 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/input is required to optimise their progress. Each school has a nominated special educational needs co-ordinator (SENCO) who should then take responsibility for implementing 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 to facilitate them.

NOTE: The way in which provision is delivered will be rather different if your child is being educated within the private system.

Placement

In children who show good recovery, mainstream education may be the most appropriate placement. For some children who show a moderate degree of recovery, mainstream schooling can be continued with adult support (for example, one-to-one help provided by a learning support assistant or '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 for any given unit, the particular focus and provision). Others may be more appropriately educated in schools

or units for children with hearing impairment. However, although in many ways, the child with an inability to understand spoken language because of LKS resembles the child with hearing loss, there are differences and these should be addressed in their educational plan.

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

Useful teaching approaches

It is essential to use strategies that allow the child's good skills to continue to develop, as these may ultimately be the way the child compensates for any residual deficits and is able to function in later life. Visual processing is usually relatively spared and can therefore be used to compensate for problems in processing auditory information and as an alternative mode of communication (see page 19 - 21).

Each child's educational programme needs to be carefully tailored to meet their particular needs. 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.

Specific patterns of impairment

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

  The Individual education plan (IEP) which is produced by the school and details the objectives for the child will need to specify ways in which pictorial and symbolic cues can be used to back up verbal explanations. Where there is a moderate or severe degree of language deficit it may also be necessary to adapt the content of schoolwork so that heavily language-based tasks or classes (for example, English) are significantly modified. It is worth noting that although number concepts are generally considered to be non-verbal, mental arithmetic (which forms a substantial and fundamental part of early years maths teaching) is a verbal skill and relies on memory and may therefore be very 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 & auditory processing**

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

  - repetition of verbal instructions several times
  - preferential seating (that is, close to the class teacher)
  - 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 work subjects that are very reliant on verbal memory (for example, history, geography)
- use of computers (supported) as the auditory requirement is minimal and there is good scope for visual cues using attractive graphics etc.

  Note: It will 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 recommendations from above will apply.

  In addition, these may also be useful:

  - a quiet and distraction-free classroom environment (as far as possible)
  - small class-sizes
  - structure the day so that tasks requiring most attention are scheduled for the time of the day when the child is most attentive (usually the morning)
  - give plenty of opportunity for positive feedback
  - ensure you have the child's attention before presenting them with a task
  - organisational prompts, for example, to pick up work-sheets, 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 periods of 'relaxing' with something the child finds easier and enjoyable (often a non-verbal task).

Key elements for a successful placement

In general, the following are some key elements in any successful school placement for a child with LKS:

- comprehensive and flexible approach giving appropriately targeted support throughout the day
- good communication between parents and school in order to capitalise on new developments in the child, and achieve consistency in management of any difficulties
- regular monitoring of the child's abilities (by speech & language therapists, educational/clinical psychologists, occupational therapists, physiotherapists and so on) and effective dissemination of this information and related recommendations or strategies, from therapists to the school and to parents
- teachers and support assistants 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
- appropriate peer group that is, a group of children with similar skills, difficulties or interests, that can provide a social network and friends
- education of the child's peers so that they have some understanding of specific difficulties and appropriate behaviour and responses. It may be helpful to use a 'Buddy' scheme to support the child.

What does the future hold? (prognosis)

Some children experience good recovery, but many are left with significant residual impairments, and it may be that there is a critical period for recovery, outside which children are left with irreparable damage. Outcome appears to be related to the length of time of the active phase of LKS. It is generally better in children with late-onset disease (language loss after the age of about 5 years), and in those with shorter periods of documented electrical status epilepticus in sleep – ESES (there is research suggesting that children with ESES lasting less than three years have better outcome). Related to this, children who respond to medical treatment of the regressions and of the ESES tend to have better prognosis, although response to treatment of the clinically visible seizures, does not generally affect outcome. In a small number of children, clinical seizures are a significant and continuing problem in their own right.

The developmental profile also has an effect on prognosis. Children who are known to have had difficulties in their early language development, prior to LKS onset, appear to have a worse outcome. LKS itself often causes difficulties in many developmental areas. Those children where the acquired difficulties are limited to language appear to do better and often respond better to medical treatment. For those children with additional acquired impairments, it is often the difficulties in social communication and interaction or general learning problems that pose the greatest barriers to recovery.

LKS may be best thought of as a spectrum, in which language tends to be first and most severely affected, but in which many other skills may be involved. Given this, 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 their progress in different skill areas over time.

The active phase of the epileptic disease typically ends around adolescence and the child's good skills, and remaining areas of difficulty should become clearer. However there is some evidence that some recovery can continue into adult life.

It is thought that in general terms, about half of the children make a reasonable recovery, a quarter have a partial recovery and a further quarter have very significant persisting difficulties.

Language outcome varies significantly. Children with a good outcome are in the minority but they usually regain competence in spoken language and tend to score within the normal range on formal assessments. Even those with good outcome however, may experience difficulties of a more subtle nature, such as problems with short-term memory and difficulties listening in the presence of noise. Those with a moderate outcome will demonstrate some degree of language impairment but spoken language will usually be their self-chosen means of communication.

Those with a poor outcome may never regain spoken language but may be able to develop skills using other communication modes such as sign language, pictures or symbols. However, because of additional difficulties with gesture and fine manipulation, signing may not be successful, and there are reports that lip-reading skills may also be difficult for the children to acquire.

Family adjustment and support

The experience of LKS is likely to be bewildering and distressing both for the child and their family. Some children may be very aware of their loss of abilities or sudden difficulties relating to their friends, and those with severe impairments of language and comprehension may find this very frightening and/or frustrating. It is not unusual for them to develop poor self-esteem and low mood as they adjust to their losses. It is important to support the child as much as possible during this difficult time, by facilitating opportunities for them to spend time with their existing friends and also creating opportunities for them to find a new and appropriate peer group, perhaps drawn from other children with language difficulties, learning difficulties, or even from the deaf community.

For parents, there is the very painful experience of having had a normal child who is apparently lost. In addition to the anxiety and distress caused by visible seizures and the need for medication or other treatments, parents must find ways to cope with a child who suddenly cannot understand the world as they did before, who may be distressed and frightened, and who may have extremely difficult behaviours and an apparent 'personality change'. Many parents report that the behavioural changes in their child, particularly aggression and sleep disturbance, are the hardest thing to deal with. As well as the demands of caring for their child with LKS, there are also the needs of any other siblings to consider, who may be bewildered and resentful of the attention paid to their brother or sister. Changes in the behaviour of a child with LKS can also directly lead to deterioration in sibling relationships and increases in fighting, another cause of family stress.

Siblings may need information about what has happened to their brother or sister, and guidance on their role, particularly as they too may have lost a close playmate and now be the target of aggression.

The course of LKS is characteristically variable and fluctuant, and the treatments are not certain, so it may be impossible to detect any steady progress in a child or to predict their future outcome, and this can be particularly discouraging for parents. LKS is a rare diagnosis and there may be little local knowledge or experience of the condition so that parents find themselves spending hours on the telephone trying to deal with local education and health services to ensure that their child's developing needs are met, or faced with a large number of differing views and approaches by successive professionals. This can be daunting, frightening and exhausting. It is common for parents to feel completely overwhelmed at times, and it is possibly all the harder that there is no identifiable 'event' such as a head injury or infection, to explain such a devastating effect on their child. It is important for parents to identify local sources of support.

Research

LKS is hard to research as the condition is rare and any centre sees relatively few children. In addition, the fluctuating nature of the disease process and seizure activity means it is hard to interpret observations. Despite this, there is a tremendous interest in this group of children, as understanding their condition would shed new light on many areas of epilepsy, language and behaviour.

There is a special interest in LKS at Great Ormond Street Hospital, and there are active plans for research into the condition.

Useful contacts

There are various sources of both practical and emotional support for parents of children with LKS, and a list of relevant organisations is given below.

FOLKS (Friends of Landau Kleffner Syndrome)
3 Stone Buildings (Ground Floor), Lincoln's Inn, London WC2A 3XL

Tel: 0870 847 0707

Website: www.friendsoflks.com
Email: info@friendsoflks.com

KIDS (Range of services provided for children with disabilities including home based learning, respite care, holiday play schemes and independent educational advisory service)

80 Wayn Flete Square, London W10 6UD

Tel: 020 8969 2817

MENCAP (support group and providers of services for people with learning disabilities)

123 Golden Lane, London EC1Y 0RT

Tel: 020 7454 0454 Fax: 020 7608 3254

Website: www.mencap.org.uk

AFASIC (UK charity representing children and young adults with communication impairments working for their inclusion in society and supporting parents and carers)

2nd Floor, 50-52 Great Sutton St, London EC1V 0DJ

Helpline 0845 355 5577 Fax 020 7251 2834

Website: www.afasic.org.uk
Email: info@afasic.org.uk

Contact-a-Family
209-211 City Road, London EC1V 1JN

Helpline: 0808 808 3555

Website: www.cafamily.org.uk
Email info@cafamily.org.uk

Epilepsy Action
New Anstey House, Gate Way Drive, Leeds LS19 7XY

Helpline 0808 8005050 Fax 0113 391 0300

Website: www.epilepsy.org.uk
Email epilepsy@epilespy.org.uk

The National Autistic Society (NAS)
393 City Road, London EC1V 1NE

Tel: 020 7833 2299 Fax: 020 7833 9666

Website: www.nas.org.uk
Email: nas@nas.org.uk

Dyspraxia Foundation
8 West Alley, Hitchin, Herts SG5 1EG

Helpline 01462 454 986 Fax 01462 455 052

Website: www.dyspraxiafoundation.org.uk
Email: dyspraxia@dyspraxiafoundation.org.uk

Hyperactive Children's Support Group
71 Whyke Lane, Chichester PO19 2LD

Tel: 01903 725 182 Fax : 01903 734 726

Website: www.hacsg.org.uk
Email : hyperactive@hacsg.org.uk

Parent Network (offers courses on parenting skills)
Room 2, Winchester House, Kennington Park,
11 Cranmer Road, London SW9 6EJ

Tel 020 7735 1214 (parent enquiry) Tel 020 7735 4596 (admin)
Fax 020 7735 4692

Skill (National Bureau for Students with Disability)
Provides information, advice and publications regarding post 16 education, training and employment for people with disability

Chapter House, 18-20 Crucifix Lane, London SE1 3JW

Information service Tel: 0800 328 5050 Text : 0800 068 2422

Website: www.skill.org.uk
Email : info@skill.org.uk

Independent Panel for Special Education Advice (IPSEA)
Provides advice and information to parents whose children have special educational needs. Professional advice for parents appealing to SEN tribunal

6 Carlow Mews, Woodbridge, Suffolk 1P12 1EA

Helpline: 0800 0184 016 Fax: 01394 380 518

Website: www.ipsea.org.uk

Department of Education and Employment (DfEE) Publications Centre
(for copies of the Code of Practice and other DfEE publications)

Tel 0845 602 2260

Advisory Centre for Education (ACE) Ltd
1b Aberdeen Studios, 22 Highbury Grove, London, N5 2DQ

Helpline: 0808 800 5793 Fax: 020 7354 9069

Website: www.ace-ed.org.uk

Makaton Vocabulary

The Makaton Vocabulary Development Project
31 Firwood Drive, Camberly, Surrey, GU15 3QD

Tel 01276 61390

Website: www.makaton.org
Email: mvdp@makaton.org

The Paget-Gorman Society
2 Dowlands Bungalows, Dowlands Lane, Smallfield, Surrey, RH6 9SD

Tel 0134 284 2308

Website: www.pgss.org

The Royal College of Speech and Language Therapists
2 White Hart Yard, London SE1 1NX

Tel: 020 7378 1200

Website: www.rcslt.org
Email: postmaster@rcslt.org

Further reading

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Commonly encountered medical concepts

LKS may also be termed:

- acquired aphasia of childhood with seizures
- epileptic aphasia
- receptive epileptic aphasia – (loss of comprehension)
- epileptic verbal auditory agnosia
- epileptic pure word deafness

Aphasia means disturbance in the ability to use language. Receptive refers to understanding or comprehension, expressive refers to use of spoken language.

Agnosia means the person is unaware of their failure to recognise, or understand.

Child development

Child development is the process by which children change and increase in their abilities in all areas (for example, motor, language, social) over time. It is viewed as a continuous process that depends on maturation of the child's brain. The brain is not fully developed at birth and grows and makes important connections, 'wiring', throughout the early years of life. Generally children follow a predictable sequence (for example, sit before they walk) although at different speeds. For the younger child, development is often assessed by considering skills in different areas such as gross motor (for example, sitting, walking), fine motor (for example, hand manipulation), vision, language, cognitive ability (for example, puzzles and problem solving) and personal-social skills. For older and more able children, it is common to concentrate on language and cognitive (non-verbal intelligence) skills.

Delay means that a child's development is not as advanced as would be expected for their age (hence it is often reported as an 'age equivalent') and this normally occurs when the child's rate of development is slower than usual.

Catch-up

Parents often think that a child can be stimulated to catch-up and then perform at the same level as other children of a similar age. This does not generally happen, as it requires development at a faster rate than normal. Most delayed children make steady progress at a slower rate than other children of the same age, and make predictable gains in learning, but never 'catch-up'.

The case for children with LKS is different. These children generally had normal early development, and were increasing their skills at the normal rate. Following a period of regression, they may well appear to 'catch-up' and learn at an increased rate, often in response to steroids. What is actually happening, however, is recovery of their previous developmental path.

Unfortunately this is not always the case in LKS, and at the end of the active phase of the disease, children or adolescents are often left with residual impairments. They may then make steady developmental progress but never regain their previous rate of learning. However there is some evidence of continuing recovery of skills into their 20's, hence they should have priority for continuing further education.

Regression is the loss of previously acquired skills, so the child appears to have returned to an earlier stage in their development. It can be uneven, and leave the child with retained isolated skills from their previous developmental level, which can mask their losses.

Epilepsy

This is a condition where a person has a series of seizures.

Seizures

These happen when part of the brain develops uncontrolled electrical activity or discharges, which stops the normal function of that part of the brain and produces the features that occur in the clinical seizure. The EEG recordings will pick up discharges and abnormalities over the area of brain affected, or even over the whole brain if the seizure becomes generalised.

In clinical seizures there is an obvious change that occurs for the person, during the seizure. This change just depends on what part of the brain is having the seizure and the person may twitch and jerk, or go blank for a few seconds or even experience a strange taste or smell.

In subclinical seizure activity there is no obvious change such as jerking, even though the EEG records electrical seizures. This does not mean the seizures are not having an effect on the person, but this effect may be on acquired skills such as language, social communication or abstract thought. In LKS, the main seizures are subclinical and occur during sleep.

Todd's paresis

This refers to temporary weakness that sometimes follows a seizure.

Convulsive status epilepticus

This is where a seizure that causes convulsions, (when the muscles of the body move out of control), continues for a long time (e.g. more than thirty minutes), or when one seizure follows another without the person regaining consciousness in-between. It is dangerous and needs urgent treatment if a seizure lasts 4-5 minutes to try to prevent a larger attack.

Non-convulsive status

This also occurs when seizures are very prolonged, or follow one upon another without break. However in

this case, the seizures do not cause convulsions but typically cause fluctuations in awareness and jerks.

Epilepsy with electrical status epilepticus during sleep (ESES)

This is a special type of non-convulsive status in which continuous discharges occupy most of sleep. It is particularly associated with the active phase of LKS and is associated with intellectual deterioration and loss of language. It may also be referred to as continuous spike and wave discharges during sleep (CSWS). This electrical activity can persist for months or even years.

Notes

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