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### **I.V.I.G. - a shot in the dark or a reasonable alternative?**

IVIG (Intravenous - gamma globulin or Intravenous Immunoglobulin) is a blood/plasma product which has been used for some considerable time by the medical profession to treat a whole variety of infection based disorders eg. Rasmussen's encephalitis and cases of primary antibody deficiency. Interest in its use in epilepsy (and to a certain extent in autism) gathered over recent years with reports of its successful use from abroad in children with LKS, LKS variant and autism.

IVIG is obtained from a large number of plasma donors and then treated to a number of processes to reduce viral transmission as far as possible. There are at least 17 different brands of IVIG commercially available and the care in its processing and its actual composition may effect both the efficacy and the amount of side effects. In very simple terms immunoglobulins are gamma shaped proteins produced by the white blood cells as part of the human immune system. The 5 different immunoglobulins are represented as IgA, IgD, IgM, IgE and IgG. Testing of the levels of each of these different groups can indicate the state of an individuals immune system and specific tests can establish allergies or whether there is a present or past infection or, an autoimmune problem or a general immune deficiency. An infusion of IVIG is a boost to the

overall immune system and may regulate an individual's autoimmune response. Immune deficiency is treated with IVIG on a 'replacement' basis but to deal with an autoimmune response a higher dose is given.

A brief look at some of the medical literature - In 1994 a study from the Netherlands 'Immunoglobulin Treatment in Epilepsy - a review of the literature' was published in 'Epilepsy' by Van Engelen et al. In a non controlled trial on both adults and children with intractable epilepsy, a significant number of the patients experienced seizure reduction and EEG improvements. In 1998 a study in Turkey of IVIG on 6 children with intractable epilepsy showed a marked improvement in 4 out of 6 cases. The Lancet in 1999 reported a controlled trial of IVIG in children with infection based obsessive-compulsive disorders including Tourettes. IVIG and plasma exchange were seen to be significantly effective in lessening the symptom severity of these disorders.

In 1996 the University of California reported that they were obtaining increasing data on the immunological abnormalities in children with autism and in some cases there was improvement of autistic features when treated with IVIG. An interesting small study in 1998 was reported in the Journal of Child Neurology on 10 autistic children with immune system abnormalities. All were treated with 4 infusions of IVIG. Out of the group 9 had only slight or no improvement to the extent that parents wished to discontinue

treatment. The symptoms in the other child improved to the extent that he moved from non verbal to speaking in short sentences and improved in sociability to the extent that he was reevaluated as non autistic. On cessation of the treatment due to the parents inability to fund more for that child there was a complete regression. A further pilot study with low dose IVIG on autistic children generally found no benefit.

Single case reports then appeared on its successful use in LKS, the most notable being the case of an 8 year old girl with Landau Kleffner Syndrome reported by Fayad, Choueiri and Mikati in 1997 'Epilepsia'. The child had developed LKS after an episode of mumps but failed to respond to anti-convulsant therapy and steroids. She was then treated with three IVIG infusions. The first two infusions produced improvements in her EEG and speech lasting for several months.

Following the final infusion the improvement was maintained without regression. The study observed that some patients with LKS may form a sub group with underlying immunologic mechanisms that respond to IVIG. Further support came from another single case study from Belgium in 1998. A boy with LKS underwent 3 phases of abrupt deterioration of language. In the first 2 phases he was successfully treated with steroid therapy but in the third phase IVIG was given instead. It was described as having 'a dramatic and comparable effect' (to steroids) in influencing language and EEG abnormalities.

These studies did not lead to any certainty about the efficacy of IVIG but rather they called for controlled studies into its use for LKS. So far no such study has been carried out. It would seem that in the U.K. at least IVIG has only been used very occasionally in LKS children as 'a shot in the dark' when other therapies are failing. In the U.S.A. it seems to be more widely given with some medical insurance companies now becoming more prepared to pay over prolonged periods for this very expensive form of treatment for children with LKS and LKS variants.

The risks - in addition to usual blood transfusion risks there are reported side effects of nose bleeds, aggressiveness and hyperactivity. Some are associated with a fast infusion rate eg. flushing, low back ache, headache, nausea, wheezing. More rarely anaphylaxis is a risk particularly for IgA depleted patients who may need to have an IgAdepleted preparation (IgAis a defence to infection and protects mucosal surfaces eg. the gut). IgA deficiency is commonly reported in autistic children. The risks however are not comparable to that involved with high dose steroid therapy.

F.O.L.K.S. are grateful to Dr. Jane El-Dahr, Associate Professor of Pediatrics, Clinical Associate Professor of Medicine and Head of the

Pediatric Allergy, Immunology and Rheumatology Section at the Tulane Hospital for Children in New Orleans for her outline of the current difficulties with research into this area:

"Since there is no data from either side of the pond, there is no answer as to what laboratory studies would be predictive of success with IVIG in children with LKS. Immunologic screening is done in the U.S. (IgA, IgM, IgG, IgG subclasses) really because if there is a deficiency of immunoglobulins the insurance MIGHT cover the cost. Low immunoglobulins do not seem to be predictive of success, however. If I had to hazard a guess the presence of brain autoantibodies would seem to be the logical place to start. Of course the difficulty is that this is not a widely available test. Unfortunately no one has looked at this type of marker (or any marker) before and after IVIG in LKS children. In the few ASD children who have had it measured prior to IVIG there seemed to be some correlation, but we don't know if the levels will drop significantly after treatment. The other immunologic parameter which has been utilised in the U.S. for treating ASD children with IVIG is an elevated titer to measles. The other difficulty with IVIG is the dose - does one use low dose or high dose? It is the high (autoimmune or 1 - 2 gm/Kg) dose which was successful in the published reports of LKS children who responded, yet what is published on ASD children has been all low dose (400 mg/Kg) dose. We need to do a good study the right way."

#### **Some parents views of IVIG -**

Linda (U.S.A.): 'My 5 year old son is successfully on an IVIG protocol... he had a set up dose of 3 consecutive days at 400mg/Kg (Baxter Gammagard). We saw immediate hyperactivity for a few days followed by great eye contact, improved behaviour and comprehension. This was outpatient, taking 7 hours the first day (could not find a vein plus a very slow rate), 4 1/2 hours the next, and 3 hours the last. He now gets 800mg/Kg when behaviours return... its been 8 weeks! His receptive language went from 18 months to 4 years. He is in regular Kindergarten with an aide and doing regular work. We are now retesting immune panels to see if we go again or wait. His expressive language is unclear but improving. The only side effect we had to deal with is hyperactivity... The immune panels are going to be matched with his pre IVIG tests. We will be looking at titers normalising, cytokines balancing... everything. He had no grossly out of whack immune problem... just when looked at as a whole it did not work.'

Ruth (U.S.A.): "Here is our experience. Prednisone - went from super hyperactive and single words to calm almost sedentary, 3 word

sentences several times /week/day. Receptive language completely normalised, except for occasional spacing out. Severe side effects from prednisone, complete regression when weaned off.

IVIG: First dose - noticeably calmer, but not so sedate. Improvement in receptive language, increase in words, but not back to the 3 word sentences. This is quite a good response considering he only got a partial dose that time.... He has continued on IVIG for 3+ years ( 400 mg /kg) - gets it every 3 months now although I can see it wearing off before we get in for the next one. Our results with IVIG: up to 4 word sentences, occasional attempts at conversation, normal receptive language, considerable improvement in social skills although still considered mod.-severe autistic. I am pleased with our results considering Jeremy was 11 before he got an LKS (probably LKS variant) diagnosis and we started this type of treatment. He just turned 16 last Saturday at least we can live with him like this and we continue to see improvement. If he had gone on like he was, we would probably, although very against our wishes, have had to institutionalise him by now, he was that bad (considered the most severely hyperactive non verbal autistic child in Portland or region at one point, now the same people are saying, "Wow! he is sure making progress ). Dr Konkol and I discussed surgery but his concern was that we could make him worse and the surgery would be irreversible and since he was showing results with IVIG, we jointly chose to continue down this path.'

Hilary (U.K.): 'Stuart had his first fit in March 1997 and his first EEG in May which was markedly abnormal. He was put on Epilim and referred to Dr. Rasmesh - a Consultant Paediatric Neurologist. Things started to go very wrong in November 1997 when he started nodding every few seconds, would lose his balance, had difficulty concentrating and was constantly exhausted. Dr. Rasmesh suggested starting lamotrigine in addition to epilim. This had no visible effect, and prednisolone was started.

From the time these multiple nodding spells had started there had been a marked deterioration in Stuart's speech and his understanding of spoken language. The prednisolone seemed to have a positive effect. He became lively, chatty, ate non-stop but his behaviour was appalling. In February 1998 the nodding, speech and language deterioration started again. A further course of prednisolone was recommended . There was a slight improvement but the nodding persisted and he was unable to go to school. I happened to mention to Dr. Rasmesh at his next appointment how jerky and twitchy he was when he was asleep. It was at this point that Landau-Kleffner was mentioned and he was referred for a sleep EEG and a MRI scan. Dr. Rasmesh also suggested that IVIG may be beneficial. Stuart

would have his treatment once a month over 3 months and he began on March 31st. The first treatment took place over 3 days, four hours a day. The following two treatments were done overnight to lessen the amount of time in hospital.

There was absolutely no difference in Stuart's condition after IVIG treatment. He started deteriorating again in June with the nodding becoming worse, he was fitting at night and seemed permanently tired. His behaviour was unpredictable, his only verbal output was screaming, he understood nothing. The only way of communicating with him was to try and sign or draw a picture. He was running away from school, climbing on roofs, locking himself in the bathroom etc. Dr. Rashmesh increased the lamotrigine to the maximum dose for Stuart. There was still no improvement and he continued on a downward slope. Dr Rasmesh's Senior Registrar then suggested clobazam. By some miracle over the first week of being on this drug (with lamotrigine and epilim) he started to speak again albeit very hesitantly but it had been so long since we heard him talk, it was just wonderful to hear the word 'Mum' again. From that date he has been relatively stable. There have been a few hiccups but he actually had a normal EEG in September 1999, his first one for 2.5 years. His speech and understanding have improved enormously and although he still has a long way to go we are trying to remain hopeful. He has settled well into the Percy Hedley School. They are fantastically supportive of both him and us and it is such a relief to see him so happy and confident. Long may it last.'

F.O.L.K.S. thank all the parents who contributed their own experiences of IVIG although many have different protocols for its administration. Some have indicated the need for prolonged periods of regular infusion to keep up improvements in language and behaviour, for others there are repeat infusions only when seizure activity returns and the IgG level is lower than normal. The expense of this treatment and lack of research into which children can benefit from it make it difficult for parent to argue for the opportunity to try IVIG therapy. Few U.K. parents of LKS children are aware of whether their child has had any immunoglobulin testing or what any blood test results were yet are very aware of their children's eczema, asthma, liability to respiratory infection, bowel problems and so on and search the health food shops for something to help. At the very least what is suggested is that in dealing with LKS we might usefully add the immunologist to the list of professionals that deal with our children.

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### Further Research Ahead - Autism and LKS

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It has been interesting to learn from Dr. Michael Chez, Paediatric Neurologist of Lake Forest, Illinois of just a few of his many research projects for autistic and epileptic children. One of the subjects for research will be the role of autoimmunity in these groups of children. The rising numbers of autistic children and the worries over DPT and MMR vaccinations have led to an urgent need to look at these issues. Studies of other autoimmune diseases eg. A.I.D.S. have led to some understanding of the role of autoimmunity even where the infection doesn't touch the brain. There have been findings of deficient IgG and IgA levels in atypical autistic children with liability to diarrhoea and ear infections which is suggestive of a functional immune system problem. In addition there is a higher frequency of serum antibodies to brain cells. Dr. Chez is suggesting that if there is an 'autistic encephalitis' it may in some cases be treatable or partially reversible. Studies with pulse doses of steroids in the U.S. shows improved responses in some 60-70% of these children compared to 10% with IVIG

Other research by Dr. Chez has been carried out on the extent of abnormal EEGs in autistic children and his findings suggest that 60% of children that regress over the age of 18 months will have an abnormal 24 hour EEG. It confirms the findings of Tuchman and Rapin of the discovery that 40 to 50% of autistic children will have abnormalities in an unmedicated 24 hour EEG compared to 24% in a one hour EEG. In particular he would suggest that the older the age at regression, especially the 24 - 36 months group the more similar to the LKS group they appear to be. Dr. Chez is also engaged in a study to establish what constitutes a normal EEG in typical young children with no family history of epilepsy or autism so that at the very least the term 'abnormalities' has more meaning.

### Other Possibilities:

**Aricept** - is an approved drug for use in the treatment of Alzheimer's disease designed to improve the functioning of the frontal lobes of the brain in the short term. A trial of the drug on Down syndrome children showed that it helped to increase spontaneous speech. Dr. Chez has conducted a preliminary study on the use of this drug in autistic and LKS children and will present his results to the American Academy of Neurology in May. He will then conduct a double blinded trial of the drug with a view to influencing the kind of apraxic, stereotypic, aggressive behaviour and poor cognitive abilities of these children. It is a drug which can cause nausea but provided the children can tolerate it,

his preliminary results are showing a positive response for some children with benefits of spontaneous speech, emotional expression, better attention and an increase in receptive and expressive language gain on this apparently low risk drug. While it is not yet known how long improvements last there appears to be some evidence that any improvements gained do not disappear when the drug is removed. It's hoped that very shortly a similar drug will be used in the trial which does not produce the nauseous side effect.

**Melatonin** - One of the many problems for families caring for LKS children is how they and their child can get a good night's sleep. Too often nights are broken to attend to seizures but sometimes it is simply that epilepsy and hyperactivity prevent a regular bedtime which leads to an increase in irritability or poor performance at school at the very least. It is also a heavy sacrifice for carers to give up their evenings as well as their days to the child. Recent studies have shown however that it may have a place for the epileptic, sleep disordered child. The manufacturer's of melatonin warn that it is not to be used for children or people with epilepsy and in some cases of very severely disabled children it has been reported as exciting seizures and it is important to obtain a medical prescription for it.

*What is it?* - melatonin is a natural hormone produced by the pineal gland. It has an ability to regulate body rhythms and promote normal sleep. It has been used in adults to help a variety of problems such as insomnia, jet lag, seasonal affective disorder, Alzheimer's disease. It is thought to work by regulating the circadian rhythm (sleep cycle) by increasing in quantity from evening to midnight and then decreasing as morning approaches. Some people have naturally low levels of melatonin such as the elderly or autistic children. In 1999 it was reported in 'Developmental Medicine and Child Neurology' as favourably influencing the sleep-wake cycle of children and adolescents.

*Its use* - its main use is to regulate sleep but in 1997 a Spanish study suggested that it could exert a neuroprotective and antiexcitotoxic effect following its use in the case of a child with severe myoclonic seizures who failed to respond to conventional anticonvulsants. It was used as 'add-on' therapy and over a period of time the seizures became controllable, resuming after it was withdrawn and stabilising when it was restored. In 1999 a study from Germany suggested that administering melatonin in the evening can exert a positive influence on the frequency of epileptic attacks in children with sleep disturbances of various etiologies. More work needs to be done to see how it can be used in intractable epilepsy.

*Side effects* - there are anecdotal reports of headache, sedation, restlessness, confusion,

nausea, tachycardia and pruritis but where pharmacologically regulated melatonin is concerned nothing other than sedation has been reported. If you are considering it you should consult your doctor for a private prescription as it is yet unlicensed. It can be relatively expensive to purchase this way. It may become less effective over time but a short break is said to quickly restore its effectiveness. Administration - melatonin comes in 3mg tablet form and is given approximately 30 minutes before bed time. It's less a case of just sedating a child but more of using it to retrain in the sleep habit so the usual routines of bath, story, teddy or whatever, go along with the tablet.

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### **'Growing Up' - teenagers coming through LKS.**

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In the last issue F.O.L.K.S. were delighted to hear from Vicki Horseywell about the progress that her daughter Claire (now 17 ) had made following the first MST (multiple subpial transection ) in the U.K. some 7 years ago. It is fascinating when the 'veteran families' of LKS children let us know of the progress their children have made despite the difficulties of the early years. Sue Webster from Stockport very kindly let us know about her son Stephen who underwent the second MST surgery and is now 17.

'Stephen has not had a fit since surgery at age 10 and his sleep deprived EEG shows minimal spiking through the night. He has been off all medication for about two years now and although it is expected that the fits recede in adolescence anyway we feel MST was the right decision to make even at 10 years old.

He attends Bladon House, part of the Honormead Group and is very happy there but we feel that this may be his last year as the level of support they can offer is not up to Stephen's ability. I cannot speak highly enough of the School for any parent who is looking for education for their LKS child. It is wonderful and has helped shape Stephen into a really nice young person.

We are currently exploring Deaf Colleges for him as we are now certain his future lies within the deaf community' (Stephen did not have a language gain from surgery and uses Makaton).

'Stephen will be 18 in April. He loves James Bond, Manchester United, his play station, his dalmatian dog Poppy, girls (although he won't admit it) and Adidas clothing. Since acquiring LKS he has been a selective eater and his range of foods and liquids is incredibly limited. He is determined to get drunk on his birthday and wants to try Vodka as someone has told him it does not taste of anything!!!!'

Sue is willing to talk to any family about 'post surgery'. We wish Stephen a very happy

eighteenth birthday and send our best wishes for his future plans.

F.O.L.K.S. would be happy to hear of other families and/or teenagers dealing with LKS. (contact 0870 8470707).

**SKILL** ( the National Bureau for Students with Disabilities) promotes opportunities for young people and adults with any kind of disability in post 16 education, training and employment. It provides information on such matters as applying to College, financial assistance, exam arrangements, what happens when an educational Statement ends and disability support takes over, training schemes and much more. The Information Service is available on 0800 328 5050 (voice) or 0800 068 2422 (text) from 1.30 - 4.30 Mon. - Fri..For anyone with a child coming towards this critical time it is a useful support service showing how to get information and advice. Much of the information can be obtained directly from its web site at [www.Skill.org.uk](http://www.Skill.org.uk). It has recently published a booklet called 'Into Higher Education 2000 "which is available by phoning 020 7450 0650. The cost is £2 to students and job seekers, for others £6.50.

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### **Computers, Language and LKS**

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Many LKS children have considerable visual ability and can work well with computers in their schools. Equally there are quite a number who resent the sheer hard work that is involved in the building of language and speech by their speech therapists nor do they understand the significance of practice. Can using specialist software overcome some of these problems? We look at a few of the very different programs that are available for children but be aware that the computer requirements are very different and some are not intended for home use but rather under the guidance of a speech therapist to monitor progress. It is important to accurately define your child's needs and discuss the use of software with your child's therapist before purchase. It is also essential to ensure the computer requirements are met adequately.

**Laureate Learning** from ROMPA - have produced a set of programs that claim to build language comprehension for a very wide range of disability and age. They are not intended to be intensive auditory training. The choice of program is dictated by where in a 7 stage linguistic hierarchy the child can be placed. At stage 1 the programs claim to be suitable for very young children or where the child has no intention to communicate but there is the beginning of a focus on the carer's speech and expression i.e. with very profound disabilities. Several programs eg 'Creature Antics' are designed to stimulate auditory attention, establish turn taking and cause and effect. The games are certainly very colourful, easy to load

(and parent and child friendly) with a sympathetic voice. There is a print out of scores obtained in the games so progress can be charted. In all the software it is essential to have an adequate size of computer or the games slow down and defeat the short attention span of some children. This range of programs in the first stages at least can be used without expressive language and less receptive language than the auditory training programs.

The stages move on with goals of improving awareness of words, developing a vocabulary, understanding categories, grammatical rules, simple sentences finally reaching stage 7 for the child of over 5 who has a developed linguistic code but needs advanced semantic training. Many parents will easily recognise in the games the speech therapy objectives that they may have had to do with pen and paper eg. voice says 'the boy is waving' and the child has to indicate he understands the object and verb. In 'Exploring First Verbs' this task is made much more interesting by presenting a colourful scene like a beach and the reward for indicating correctly is an animated character. Most of the programs can be rerun at different levels to vary the complexity. 'Nouns and Sounds' is a program to encourage listening and sound discrimination offering seven different activities. A useful feature is that this Company sells a Magic Touch screen to allow for interacting by just touching the screen with the finger for those children who have very poor mouse control.

The prices vary per program, ranging (for one copy) from £75 to £169 and there are different programs in each stage which are exclusive so more than one may be required. Recommended computer requirements - Microsoft Windows '98, Pentium P100 CPU, hard drive, windows compatible sound card and speakers, 4X speed CD-ROM, 16MB RAM or above. A free demonstration CD-ROM is available. This and other information can be obtained from [www.laureatelearning.co.uk](http://www.laureatelearning.co.uk) or Laureate Learning, Goyt Side Road, Chesterfield, Derbyshire, Tel 0845 3000899

**Fast Forward** 1 and 11 (from Scientific Learning) - these are intensive CD-ROM auditory training programs for children with language and reading problems. The program consists of a set of 7 computer games carried out under the supervision of Fast Forward trained providers in schools, clinics etc. It can be done at home with an Internet facility but is therefore a very expensive course to take. This is required by Fast Forward as the data from the child needs to be analysed and recorded. The child has to attend for 100 minutes per day, 5 days per week, for 4 - 8 weeks. This may not be suitable for every child with a language problem but it claims to improve auditory processing, phonological awareness and stimulate language development by training the ability to distinguish the various

components of speech. Certainly some LKS children would find this very demanding if not impossible during periods of regression but possibly very necessary in less severe phases. No expressive language is required but quite a lot of receptive language would be needed to follow the instructions within the game.

The games are colourful and animated and reward success. 'Phonic Words' - two colourful pictures representing a similar sound eg bee/knee. The child has to choose the picture on hearing the word which is given very slowly at first and then as progress is gained more quickly.

'Circus Sequence' - is used to train recognition of non verbal tones. Old MacDonald's Flying Farm - a child is asked to identify the change of sound in a series of repetitive sounds during the course of a computer game of catching animals. 'Language Comprehension Builder' - a set of colourful pictures accompanied by a sentence. The child matches the meaning to the correct picture. 'Block Commander' - a game where the child follows increasingly complex commands to increase comprehension and attention. 'Phoneme identification' - an animated character gives a phoneme, two versions are then given and the child has to match the correct one and finally 'Phonic match' - a game designed to stimulate word recognition and auditory memory.

It requires a child to attend for a full 20 minutes at a time otherwise progress could be slow. The idea behind auditory training is simply that the brain has to be trained to efficiently make the connections between different sounds and training or retraining the brain takes a lot of effort.

Further information can be obtained from [www.scientificlearning.com](http://www.scientificlearning.com)

**Earobics** by Cognitive Concepts- again this is an auditory training program which claims to focus a child's attention on sounds and teaches phonological awareness, auditory processing and language comprehension with the general aim of improving reading and spelling skills. They state that all children benefit (and especially in developing reading skills) from the game but it can be used for children with special learning needs. It is available as Earobics 1 (normal developmental ages 4 - 7 but not necessarily skill level), Earobics 2 (normal developmental age 7 - 10.) and also Earobics for adolescents and adults. They recommend 15 - 20 minutes per day but state that children with special needs may need more. It certainly does not have the intensity of Fast Forward but unlike Fast Forward there is a Home version (for 2 players) and other more sophisticated versions such as the Classroom, Pro Plus and Clinician versions. They all use the same games but the Classroom and specialist version have features for producing reports and data tracking and a greater number of players. It is intended that teachers would use it within their

own program and customise it. They are not available to parents.

Earobics 1 has 6 interactive games with 309 levels of play eg. Karloons balloons (38 levels of play). The child has to save coloured balloons for a clown by correctly remembering the order of sounds they hear. The reward for correct identification in this case is that the balloons don't pop. C.C. Coal Car (74 levels of play) is a game where a child need to identify long and short vowels and consonant sounds, in isolation and then in a word. The reward is the coal car is loaded. In Caterpillar Connection (56 levels) when a child blends sounds into words correctly a caterpillar changes into a butterfly.

Earobics 2 (593) levels of play in 5 games practices the same skills but at a more advanced level in such games as 'Calling all Engines'. This is designed to improve auditory sequential memory and to follow directions against an increasing background noise. A game which many LKS children could find very difficult. Other games develop rhyming skills, spelling patterns etc.

Further information from [www.earobics.com](http://www.earobics.com)  
Minimum requirements: Windows, 4486/66MHz or faster, Windows 95/98/NT Pentium or faster, 24 Available RAM 3MB, available hard disc 640x480x256 colour display SoundblasterTM - compatible 16 bit sound card and speakers  
Macintosh - Apple Macintosh 68040 or any power Pc 7.1 or higher, 24MB available RAM, 3MB available hard disc space 640x480x256 colour display 4 x CD-ROM drive, SoundblasterTM - compatible 16 bit sound card and speakers.

It may be that your children are using different programs or are benefiting in other areas from using computer software. Do write to us and share your experiences with other families in FOLKS.

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### **'Learning from experience'**

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Some of the most useful and extensive information about L.K.S. comes from the parents, carers and families of children with L.K.S. We were grateful to member Gary Cheshire for imparting that information and experience in a presentation to The Jane Moore Trust in November.

Gary and his wife Ilene became the carers of an LKS child just over a year ago and had to set about meeting the challenge of L.K.S. for the first time. They presented their experience to an audience of different professionals working in the field of child care. Gary described the syndrome, it's history and the battles and rewards of meeting the needs and demands of such a child.

The presentation was well received and the audience were particularly interested to hear of

the obstacles that families face in obtaining accurate diagnosis and the importance of the passage of information between everyone concerned with a child's welfare.

The Jane Moore Trust - is a family placement service based in West Lothian. It places and supports children in specialised foster care with a multi disciplinary professional team.

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### **A Parent's Story: Richard Eiffert**

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Richard was born in Kingston Hospital, Surrey in September 1990, weighing 10lb. 5oz, very big and very jaundiced. The first six weeks were trouble free, then came his first chest infection, then ear infection. He was to alternate between the two for the next two years. Life was a constant round of doctors visits and antibiotics. He remained surprisingly, a happy and good natured child.



We moved to Suffolk when our other son David was 6 and Richard was 2 years and 6 months. By which time his speech was coming along slowly. Shortly after this Richard was given his Hib (Haemophilus Influenzae B) vaccine. The next day he threw his first ever 'tantrum'. It lasted an hour with him screaming and throwing himself backwards and forwards in a buggy (we were collecting David from school) so much that the pram broke in half. A complete shock. Strangely we did not connect this to the Hib until many years later.



Richard's speech remained slower than average and a bit confused at times. He was referred to a hearing clinic and a speech therapist who thought I was an 'over protective mother' comparing him to his older brother.

Richard started mainstream school in 1995. The so-called speech therapist had by now discharged him because he was 'fine.' The year went quite well, he was counting to 10, had a basic knowledge of the alphabet and was reading simple books. The school year ended on July 24th 1996.

On July 26th at 3.30 a.m. our whole world was to change. We were woken by the terrifying sound and sight of Richard having his first epileptic seizure. A one off, our GP assured us when he arrived! By 6.30 a.m. the same morning he was admitted to Ipswich hospital after his 2nd seizure.

From then until March 1997 we learnt to deal

with epilepsy and accept that Richard had some learning difficulties. He was now on a ROS 2 (Record of Support) at school with 1-1 supervision at lunch and play times.

In late February Richard was given his MMR booster after which we were to see a devastating decline in him. His epilepsy became so much worse, all types of fits, he had to wear a protective helmet, his behaviour was uncontrollable and his speech went down to 5 words. He was so confused and frightened, just like us. The education authority allowed Richard to only attend school with a 1-1 for 15 hours a week plus 5 hours home tuition. He was often too ill and weak and badly behaved to attend any of those at all. It was a sad and harrowing time.

Many tests were carried out at Ipswich Hospital, all of which proved negative. There seemed to be no answers to his regression. Flicking through a book on epilepsy I came across Landau Kleffner Syndrome under the heading 'disabling conditions'. I knew immediately that this was what he had. We insisted on a sleep EEG which was carried out at the Park Hospital in Oxford and LKS was confirmed. We were transferred to Great Ormond Street Hospital in October 1997 and Richard was started on Prednisolone corticosteroid treatment. Fortunately these had a great impact on Richard, and 10 days later his seizures stopped, and by Christmas his words were returning. The steroids were pulsed after the first 3 months and Richard was to remain on them for the next two years,



until December 1999. During this time his condition fluctuated, in terms of the epilepsy his behaviour and his speech and language, but the improvement has been tremendous.

In June 1998 we moved to Lincoln and Richard started at I CAN's Dawn House School in Nottinghamshire. A day pupil in a speech and language school. He has progressed a lot in which we feel is an appropriate school environment for Richard. An encouraging and speech orientated curriculum in a structured and calm atmosphere.

Richard has also been on a gluten and casein free diet for nearly a year, since April 1999. This is based on the investigations into the theory that autism, and autistic spectrum disorders, could be a consequence of undigested proteins leaking into the blood stream, due to a more permeable gut, which itself could be caused by the over use of antibiotics and/or MMR or other childhood vaccination, causing damage to the brain. We

feel that we have seen an improvement in behaviour and seizure control, however we have no medical proof and are carrying this out independently of Great Ormond Street Hospital.

Richard has recently had a Spect scan at Great Ormond Street Hospital and further tests are being carried out. MST surgery may still be a possibility for him in the future. His behaviour is manageable now but still unpredictable, he still has seizures but not as frequent, and we have no idea if he will regress again to his pre steroid state. Although he has quite a lot of speech, Richard still has major comprehension and word finding difficulties, and he is still unable to read which he finds very frustrating. Not knowing is very difficult, the worry about the future always there. We long for our life before LKS for Richard, for us and for David.

We still have high hopes for Richard and only time will tell if he can fulfil his dreams. That of any typical 9 year old!!!!..to be a lorry driver or a policeman!

DIANE and FRED EIFFERT

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### **'Challenging Behaviour - on the receiving end'**

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Some members may have watched the T.V. drama '*Kid in the Corner*' which portrayed what it is like to live with a child with ADHD (attention deficit hyperactivity disorder). Committee member Hilary Heron spoke for many parents when she wrote to the Radio Times who published her letter:

'Having watched '*Kid in the Corner*', I felt compelled to congratulate Channel 4 and Tony Marchant. As the mother of a so-called 'difficult child' - a boy - who suffers from a 'syndrome' that hardly anybody has heard of, it was at times too painful to watch. I, too, at times have wished my son had never been born. We too have experienced prejudice, misunderstanding, embarrassment and total despair. Yet the joy he can bring far outweighs any of this. Those people whose tolerance of children with less than perfect behaviour is non-existent should be encouraged to watch this and perhaps keep their cruel comments and lack of understanding to themselves'.

FOLKS are grateful to Megan Hawkes for her very moving account of the pressures upon parents that the public perception of children's behaviour (often very ill children) can cause:

#### *'Peace on Earth, Good Will to All'*

'The holiday season... oh what fun to take a 5 year old and a 3 year old to Walmart on a rainy Saturday.!

I actually ended up there because my car would not start. Of course, batteries only die on cold rainy days, and this was another expense

that was really not on the top of my wish list. But here I was, getting more and more frustrated as I stood in line waiting to talk to the sales person who was more interested in what the person on the phone was saying than in the customer in front of them. My two children sat crammed together in the blue shopping cart that I didn't dare let them out of. Mommy! I WANT THAT SMILEY FACE BALLOON! SHE TOUCHED ME! SHE PULLED MYHAIR. and loudest of all, 'YOU SAID IT'S MYTURN TO HOLD MOMMY'S BOX OF TAMPONS!!!!'

But I have parenting skills, and I was by gosh gonna use them. I tried to remain full of grace and composed. So I turned to my beloved offspring and said, "Girls, you are both about to start losing privileges. Now, behave. I am finding your behaviour unacceptable." Not a hint of red around my cheeks either, as I said it. Ok, well, that might be stretching the tale just a bit. Inside, I was more than just a bit irritable, but I was rather proud of my outward composure. Did I mention what personal product Amanda and Heather were fighting over? You can probably figure out the rest of my mental state.

But the best was yet to come. While I waited for the salesgirl to quit popping her gum and hang up, a charming young man in line behind me spoke loudly to his companion, "Man, if that wuz MYkid, I'd take them out back and slap the (expletive) outa them. I didn't never get away with no crap like that when I was a kid. My ol' man would a busted my butt." He and his companion went on to stare rudely and roll their eyes as I continued to attempt to quiet my daughters.

I thought, "Take a big breath. You can deal with that." But how do you deal with that?

In spite of my red face and rising anger, I held on. After all, the person behind me couldn't have any idea of what we've been through the last few years. They couldn't know the fears we've had of losing our kids, of seizures that take on a nightmarish quality all their own at 4:00 a.m.

They couldn't know about the rollercoaster rides of new emotions and RAW emotions that we've been on since our daughter begun the journey of paediatric specialists and the quest to define and control my daughter's illness. They couldn't be aware of all the uncontrollable tantrums that turn into seizures, tremendous weight gain from medications, and the feeling that accompanies watching your child turn blue in an ambulance.

They must not know about seeing your baby unconscious, and of pacing back and forth in an emergency room as you waited forever to find out what was happening to her.

Nope. They have every right to stand their and call out their judgement upon others. What they see is a chubby little kid whining, crying and fighting with her sister. What they don't see is a

child, frustrated by the way she feels physically, overly stimulated by the endless greed-fest that society has turned the holidays into, who is unable to control her emotions because of new medications. What they don't see is a kindergartener who is chronically exhausted, but smart, enough to know that she's different. How would they understand about a child who at only 5 years old is dealing with body image problems from weight gain and clinical depression as a side effect from new medications?

And they sure couldn't understand that this is a little girl who at times in her life has screamed and cried when she's just seen people wearing white coats. How would they know that I really didn't want to be at Wal-Mart at that moment? Or, in fact, that going anywhere is a chore, from the moment the 45 minute battle begins where I have to force my daughter to get her own shoes on. They couldn't understand that if it was really my choice, the last thing I'd want to do is leave home at all with my kids, and that often as not, I feel imprisoned by my own children's physical 'issues'.

But most of all, they couldn't see how much it hurt me to get to be a victim of ignorance and intolerance. It would have been worse for me if I had actually cried before I got back to the car.

After all, I don't take my kid out back and slap them around - although I can guarantee the urge does occasionally hit me. In fact the urge to reach out and slap someone landed on me pretty hard at Wal-Mart at that moment, but it sure wasn't my kids I was tempted to slap.

In the faith that my family practices, we believe that God wants us to turn the other cheek. At that moment those instructions felt a pretty big order. What I wanted to do just then and what I will do are two different things. What I really will do, is pray for my kids. I'll pray that as they are more and more out in the world, that they will not run into intolerance and ignorance all of their lives, like that which fell from the lips of the outspoken gentlemen in line behind me.

And while I'm at it, I will pray for peace on earth and good will to men. And women and children at Walmart.'

'Challenging behaviour is defined as 'behaviour of such intensity, frequency or duration that the physical safety of the person or others is likely to be placed in serious jeopardy, or behaviour which is likely to seriously limit or deny access to and use of ordinary community facilities or impair a child's personal growth, development or family life.' - The Mental Health Foundation (1996). There are no easy answers to the problems of challenging and destructive behaviour of children with LKS. In those periods when it 'just has to be lived with' parents need to know they are doing what is right and doing what they can. FOLKS are grateful to Sherry

Humphreys of Edmonton, Alberta for her recommendation of the book 'Secrets of Discipline- For Parents and Teachers' by Ronald G. Morrish ( 1997/98 Woodstream Publishing, Ontario, Canada). There is also a topical booklet available from the Mental Health Foundation, 'Legal Issues arising from the care and control of children with learning disabilities who also present severe challenging behaviour. A Guide for Parents and Carers'. The cost is £4.50 and can be obtained from 20/21 Cornwall Terrace London NW1 4QL( tel: 0171 535 7400).

The Challenging Behaviour Foundation is a Charity which amongst other things exists to provide information to parents and carers. Leaflets on such topics as 'Physical Intervention', 'Basic Information', 'Who Can Help', 'Communication', and 'Safety Adaptations' are available. There is no Helpline as such but obtaining a leaflet allows an opportunity to discuss challenging behaviour issues and to obtain a regular newsletter. An explanatory leaflet can be obtained by writing to The Challenging Behaviour Foundation, 32 Twydall Lane, Gillingham, Kent. ME8 6HX (tel/fax: 01634 302207).

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### **'Making Magic'**

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F.O.L.K.S. is not quite sure how member Maggie McMahon managed to successfully stage a really enjoyable and entertaining event at The Grange Hotel, Holborn, London on 24th January in just 10 weeks or so but she certainly did. In the last issue Maggie circulated the story of her LKS child Melissa in order to make the public more aware of the heartbreak that can go along with the label LKS. She then joined forces with her friend Patricia Rogerson, an events organiser and they set out to make it happen. Maggie and Patricia's talent and determination produced a sparkling event at which 120 people attended and were treated to an excellent buffet supper at a luxurious hotel in central London. The evening was packed with musical entertainment from artists who freely gave their services. A Charity Auction was held which included such items as Paul Scholes football shirt, tea for two at the Dorchester and a supper party for 8. Guest heard a little of what FOLKS does as a parent group and what it is like to be a parent with an LKS child. As a result of Maggie and Patricia's hard work and the generosity of those who attended, funds were raised for both F.O.L.K.S. and St Elizabeth's School in Hertford which Melissa attends. FOLKS received over £2,000 from the event and also £400 in donations which will go towards the FOLKS DAY 2000 for parents, professionals and children.

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### **Christmas Carols in the Cold**

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The last Christmas of the old Millenium was given its true meaning in Reading due to the kind hearts and beautiful voices of 'The Reading Bach Choir'. Member Janet Sunman nominated F.O.L.K.S. as the Charity for which the Choir would sing. Choir member Lesley Boardly and family friend of Janet, says:

'You will be pleased to know that after an hour's singing by 24 people (with 2 children 'minding' the collection buckets) £140 was collected. We hope the money may help a little and perhaps even the publicity at the station may have alerted people to the syndrome. It must be very hard work to keep such a group as yours going when you have children to care for with above average needs. All the best with your endeavours.

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### **USEFUL SOURCES:**

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#### **'Apraxia Kids'**

We are grateful to FOLKS member Marie Riddle for letting us know about the existence of an Internet site called 'Apraxia Kids'. This group started life as an e-mail group for parents in the U.S.A. but expanded rapidly to serve parents and professionals all over the globe. Apraxia (often used interchangeably with the term 'dyspraxia'). Praxis is the ability to execute skilled movement, a child with apraxia has difficulty in sequencing and executing speech. The common characteristics may also include difficulty with reading and spelling, fine motor movements and sensory integration difficulties.

Where LKS children have this particular kind of motor speech disorder as a feature of their condition, the web site of this group contains very helpful information for families. Among the topics covered are how to obtain a speech and language evaluation, literacy development for children with apraxia, how parents can help at home, and a variety of speech and language topics.

Apart from the information on the site (<http://www.apraxia-kids.org/aksubscribe.html>), the e-mail group is free to join and is very active. To subscribe to the group e-mail [listserv@listserv.syr.edu](mailto:listserv@listserv.syr.edu) and in the message portion type subscribe apraxia-kids. Details of local parent groups in the U.S.A. and Australia are available on the site. For those parents without e-mail in the U.K., similar information and support is available from the Dyspraxia Trust, 8 West Alley, Hitchin, Herts. SG5 1EG. Any member needing to know more about 'Apraxia Kids' but does not have e-mail should contact F.O.L.K.S. Helpline -0870 8470707.

Our thanks to Folks Lister Jill who told us about a useful internet site for American Sign Language with animated signs at [dww.deafworldweb.org/sl/](http://dww.deafworldweb.org/sl/) and also to Fred Eiffert for directing us to the alternative Paget Gorman site at <http://www.pgss.org/>

#### **'The National Society for Epilepsy'**

The Society has a number of different services apart from their main object of providing for the assessment, treatment and care of adults with epilepsy particularly in the south east of England. They also provide information and education services for all ages. Their confidential Helpline (on 01494 601400) is available between 10 a.m. and 4 p.m. from Monday to Friday. They say that 30% of their Helpline work deals with calls from parents of children with epilepsy. They have a training department to train organisations such as schools and carers of children with epilepsy awareness and the administration of emergency medication. A series of leaflets are available from the Society at The Chalfont Centre for Epilepsy, Chalfont St. Peter, Bucks SL9 0RJ. In March the first phase of their new web site will be launched which contains more detailed information than is contained in the leaflets. The site will be [www.epilepsynse.org.uk](http://www.epilepsynse.org.uk).

#### **'The Epilepsy Association of Scotland'**

The Association provides a wide range of services including a National Helpline, literature on all aspects of epilepsy, local branches throughout Scotland, an Epilepsy Resource Centre, training in managing epilepsy to professional groups, community support for adults with complex needs, liaison within Epilepsy Clinics, an advocacy service, advice, support and counselling.

Address: 48, Govan Road, Glasgow. G5 1JL  
The Helpline: 0141 427 5225 is available Mon - Fri 9.00a.m. to 4.30p.m. Website - <http://www.epilepsyscotland.org.uk/epilepsy>.  
E-mail - [admin@epilepsyscotland.org.uk](mailto:admin@epilepsyscotland.org.uk)

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#### **FURTHER FROM F.O.L.K.S.**

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**Helping others in a different language** - one of the hardest tasks of the last 12 months has been trying to deal with requests for help and information by parents who have no English. Mr. Gerald Hantusch spent over a month 'sweating blood' to translate parts of the Information Pack into German. He succeeded (with the 'assistance' of a machine translation) and we have sent out our first Pack out in German. It is also now on the FOLKS web site via a link. We are indebted to him for his efforts and for starting work on the French version. Translation is almost complete on a Spanish

version. If any of our members can help or know of someone who can help with the occasional short translation into another language (in particular French, German, Italian, Spanish, Urdu or Punjabi) we would be very glad to hear from them.

**Running for FOLKS** - on April 16th. Barbara Wilson will undertake the London Marathon as a sponsored run for FOLKS. The major charities are often represented in the Marathon and this is the first year that anyone has run for FOLKS. It will be her first marathon, we wish her the very best of luck, good weather and not too many blisters.



**Sharing experiences** -when member Beverly. Williams sent FOLKS a Christmas card we suddenly woke up to the fact that there were a number of parents in the Leicester area that could actually meet, get together for coffee and 'LKS children discussion'. So we did. With nine of us it almost became a FOLKS branch meeting at the Stakis Country House Hotel. For over 3 hours we exchanged views and information on LKS.. An odd thing about LKS that while our children are all very different the parents have so many experiences in common. We enjoyed it so much we are going to meet again soon. Anyone interested in joining us contact 0870 8470707

A similar meeting became a possibility in Essex/Herts. and we met with 7 members of FOLKS in Basildon and then again in Beds./Herts It's astonishing that in meeting other parents there is such an amount of detailed information and experience that you can tap which is unavailable from any other source. No 20 minute appointment with a doctor however well qualified or any textbook can give you this kind of insight into something like LKS. Thanks to everybody who made the effort to come.

The Committee met again in January to get down to dealing with the logistics of **F.O.L.K.S. DAY 2000** on 3rd. June in yet another new venue by the kind invitation of St. Elizabeth's School, Much Hadham, Herts.

We are very pleased to welcome our Honorary member Professor Brian Neville and Dr. Andrew Holton, Consultant Paediatrician of Leicester Royal Infirmary as our speakers. The afternoon will be divided into a series of workshops on 'Learning with LKS' and 'Communication' (with Mary Jennings, Senior Speech and Language Therapist of the John Horniman School). Families are free to visit either workshop during the course of the afternoon. Many activities are planned for the children

throughout the day which with the help of 'The Leisure Team' who coordinate and supervise fun activities particularly for children with different disabilities and the Hertfordshire Red Cross. We look forward to seeing you.

**If you haven't done so already make sure your registration forms are completed and returned. It will help us to organise the children's care more effectively if you can return your forms as soon as possible and by the 12th. May**

F.O.L.K.S.NEWS welcomes all contributions, articles, letters and comments for publication. If you have any item suitable for publication it should be sent to **PO Box 749, Erith, Kent DA8 3AU** by 1st September 2000. F.O.L.K.S. does however reserve the right to omit or edit items where necessary. F.O.L.K.S. NEWS is published to provide general information to parents and carers of children with Landau Kleffner Syndrome and to interested professionals.

The contents are not and are not intended to be, a substitute for advice from a qualified medical practitioner, preferably one experienced in the management of this complex disorder.



# F.O.L.K.S. Data Sheet

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Surname \_\_\_\_\_ LKS Child's Name \_\_\_\_\_

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Parents Forenames \_\_\_\_\_ LKS Child's Date of Birth \_\_\_\_\_

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Siblings Name(s) and Age(s) \_\_\_\_\_

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Address \_\_\_\_\_

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Country \_\_\_\_\_

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Tel No(Home) \_\_\_\_\_ (Work) \_\_\_\_\_ Fax No \_\_\_\_\_

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Best Contact Time \_\_\_\_\_ E-Mail Address \_\_\_\_\_

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Doctors Names \_\_\_\_\_

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Hospital \_\_\_\_\_

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Age of LKS Onset \_\_\_\_\_ Age of LKS \_\_\_\_\_

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Diagnosis \_\_\_\_\_

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## Seizures:

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Type of Seizures \_\_\_\_\_

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Frequency of Seizures \_\_\_\_\_

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Suspected Causes: <small>(please specify)</small>	<b>Birth Related</b>	<b>Viral Related</b>	<b>Vaccine Related</b>	<b>Other</b>
Behaviour Problems:	<b>Mild</b>	<b>Moderate</b>	<b>Severe</b>	<b>Very Severe</b>
Communication Problems:	<b>Mild</b>	<b>Moderate</b>	<b>Severe</b>	<b>Very Severe</b>
Speech Deficit:	<b>Mild</b>	<b>Moderate</b>	<b>Severe</b>	<b>Very Severe</b>
Motor Difficulties:	<b>Mild</b>	<b>Moderate</b>	<b>Severe</b>	<b>Very Severe</b>

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Current medications \_\_\_\_\_

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Past Medications \_\_\_\_\_

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\_\_\_\_\_

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Surgery/Other Treatments \_\_\_\_\_

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## Education:

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School Name \_\_\_\_\_

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School Type \_\_\_\_\_

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Speech therapy **Y/N** (Details) \_\_\_\_\_

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Adult Support **Y/N** (Details) \_\_\_\_\_

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LEA Name (UK Only) \_\_\_\_\_

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Statemented (UK Only) **Y/N** \_\_\_\_\_

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If you consent to the release of your Name, Address, Tel/Fax Nos., and E-Mail address to other LKS Parents please tick this Box.