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# Landau-Kleffner Syndrome: Treatment Options for Epileptic Manifestations and Language Rehabilitation

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## 1) Introduction

Landau-Kleffner Syndrome (LKS) was first recognized by William Landau and Frank Kleffner in 1957 (Fandino, Conolly, Usher, Palm, & Kozak, 2011). Initially, children with this syndrome were mistakenly thought to be either deaf, or on the autism spectrum. This rare syndrome is a form of acquired childhood epileptic aphasia usually occurring in children between the ages of 3 and10 years old. (Fandino et. al., 2011). Seizures, EEG abnormalities, auditory agnosia, and a loss of language all accompany this syndrome. Few theories exist as to what causes LKS; however, to this day no concrete etiology is known (Kuriakose, et. al., 2012).

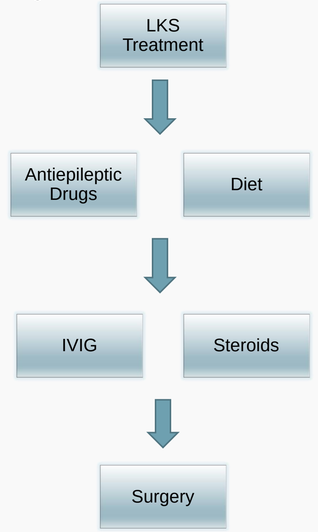
## 2) Important Characteristics

- Electroencephalographic abnormalities are present primarily in the temporal lobe (Mohamad & Shamseddine, 2005).
- More predominate in males than females (Mohamad & Shamseddine, 2005).
- Seizure activity present in approximately 80% of cases (Mohamad & Shamseddine, 2005).
- Deterioration of previously normal developing language skills (Morrell, 1995).
- Auditory processing difficulties (Fandino, 2011).
- Associated behavior problems such as hyperactivity (Mohamad & Shamseddine, 2005).

Table 1

| Treatment | Pros | Cons |
|---|---|---|
| Antiepileptic Drugs | Proven effective in most cases. | Does not prove to positively effect language. |
| Ketogenic Diet | Lasting impact on improving seizures and language functioning. | Very limited information exists regarding efficacy of this treatment. |
| IVIG | No known adverse side effects. | Only shows dramatic improvement in select LKS patients. |
| Steroids | Rapid recovery of language has been noted in some cases. | May not yield any results due to fluctuating nature of LKS. |
| Surgery | Effective in most patients after other treatment methods have failed. | Most invasive form of treatment |

Figure 1. Treatment Course for LKS




## 3) Treatments

There are different treatment options for managing the seizure activity in children with LKS. Mohamad and Shamseddine described the various options (2005).

1. Antiepileptic drugs: valproic acid, diazepam, ethosuximide, and clobazam, among others.
2. Ketogenic diet: a high-fat, adequate-protein, low-carbohydrate diet. Traditionally, glucose fuels the body in a high carbohydrate diet. However, this nutritional scheme forces the body to burn fats rather than carbohydrates. As a result, instead of converting carbohydrates into fuel for the brain the liver will convert fat into fatty acids and ketone bodies. The ketone bodies replace the glucose in the brain.
3. Intravenous Immunoglobulin (IVIG) : a blood product containing polyvalent IgG antibodies is administered intravenously.
4. Steroid treatment: corticosteroids, specifically oral corticosteroids are most often used.
5. Surgery: multiple subpial transection (MST). This technique was first described by Frank Morrell, this surgical method involves severing the horizontal connections while maintaining the vertical connections in the brain where the electroencephalographic abnormalities are most present (Morrell, 1995).

## 4) Language Rehabilitation

After seizure management, most patients with LKS do not regain their language function. Additionally, patients develop an auditory agnosia; thus, alternative means of communication must be implemented in order for patients to regain loss of language function (Mohamad & Shamseddine, 2005). Different methods by which language rehabilitation can occur are as follows: speech therapy, sign language, color coding, visual cues, etc. (Malvestio, 2010).

The two most important methods of treatment as described by Malvestio are speech therapy and sign language usage. (Malvestio, 2010).

## 5) Why Sign Language?

- The use of sign language stimulates not only the right hemisphere, but the left hemisphere as well (Gordon, 2003).
- By using sign language , children with LKS are able to process information bypassing the auditory pathway which has been rendered incapable of processing verbal stimuli. (Perez et. al., 2001).
- Sign language may facilitate the recovery of oral language because using sign language stimulates functionally connected core language networks (Deonna, et al., 2009).
- Additionally, using sign language as a means of communication will not only improve the child's communication abilities but also alleviate anxiety and improve social skills. (Malvestio, 2010). Thus, the child will also have the ability to gain access to a linguistic and social community while developing their cognitive function (Woll, 1996).
- Many children are able to develop a proficiency in sign language comparable to that of other children who use sign language as their primary mode of communication (Perez, et. al., 2001).

## 6) Conclusions

•We currently understand the development and the characteristics of the syndrome better than we did 20 years ago.

•Long term outcomes for patients with LKS in terms of language rehabilitation and EEG normalization are variable.

•There is no ideal treatment option and controlled clinical trials are needed in order to assess the best method of treatment for EEG abnormalities.

•Sign language and speech therapy seem to be the best treatment options for language recovery; however, more research is needed.

•Unfortunately, due to the rarity of the syndrome, it is difficult to assess long term outcomes and create controlled clinical trials with these patients.

## References

Deonna, T., Peter, C., & Ziegler, A. L. (1989). Adult follow-up of the acquired aphasia-epilepsy syndrome in childhood. Report of 7 cases. Neuropediatrics, 20(3), 132-138. doi: 10.1055/s-2008-1071278

Deonna, T., Prelaz-Girod, A., Mayor-Dubois, C., & Roulet-Perez, E. (2009). Sign language in Landau-Kleffner syndrome. *Epilepsia, 50*(6), 777-782.doi:10.1111/j.1528-1167.2009.02226.x

Fandiño, M., Connolly, M., Usher, L., Palm, S., & Kozak, F. K. (2011). Landau–Kleffner syndrome: A rare Auditory Processing Disorder; Series of cases and review of the literature. *International Journal of Pediatric Otorhinolaryngology, 75*, 33-38. doi:10.1016/j.ijporl.2010.10.001

Gordon, N. (2004). The neurology of sign language. *Brain and Development, 26*(3), 146–150. doi:10.1016/S0387-7604(03)00128-1

Kuriakose, S., Lang, R., Boyer, K., Lee, A., & Lancioni, G. (2012). Rehabilitation issues in Landau-Kleffner syndrome. *Developmental Neurorehabilitation, 15*(5), 317-321. doi:10.3109/17518423.2012.701241

Malvestio, C. (2010). Landau-Kleffner Syndrome (LKS): A rare childhood neurological syndrome. *American Journal of Electroneurodiagnostic Technology, 50*(2), 122-132.

Mikati, M. A., & Shamseddine, A. N. (2005). Management of Landau-Kleffner syndrome. *Pediatric Drugs, 7*(6), 377-389. doi:1174-5878/05/0006-0377

Morrell, F., Whisler, W. W., Smith, M. C., Hoeppner, T. J., de Toledo-Morrell, L., Pierre-Louis, S. J.,... Bergen, D. (1995). Landau-Kleffner syndrome. Treatment with subpial intracortical transection. *Brain: A Journal of Neurology, 118*(Pt 6)1529-1546. doi:http://dx.doi.org/10.1093/brain/118.6.1529

Perez, E. R., Prélaz, A., Metz-Lutz, M., Braem, P. B., Deonna, T. (2001). Sign language in childhood epileptic aphasia (Landau-Kleffner syndrome). *Developmental Medicine & Child Neurology, 43*(11), 739-744. doi:10.1111/j.1469-8749.2001.tb00154.x

Woll, B., & Sieratzki, J. S. (1996). Sign language for children with acquired aphasia. *Journal of Child Neurology, 11*(4) 347-348. doi: 10.1177/088307389601100419