

50 years Landau-Kleffner Syndrom: A look back and an outlook to the future. An interview with Dr. William M. Landau

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My presentation is about the Landau-Kleffner syndrome (LKS).

I am going to answer questions about this disorder based on an interview I conducted with Dr. William M. Landau in St. Louis Missouri on October 21st 2005.

Terminology

The neurologist Landau and the educationalist Kleffner described in an interdisciplinary work the disorder by describing six cases.

In literature different synonyms are used:

1. Landau-Kleffner syndrome after Landau and Kleffner who were the first to describe this disorder.
2. "Syndrome of acquired aphasia with convulsive disorder" is the title of the original work which appeared in the magazine "Neurology" in 1957.
3. "Verbal auditory agnosia in children" was issued in Isabelle Rapin's publication in 1977.

The terminology reflects the confusion and ambiguity which occurs around this syndrome. So the root of this problem are different theoretic views.

William Landau's publication in *Archives of Neurology* defines the label "Landau- Kleffner Syndrome" as "an eponymic badge of ignorance".

Still it is more useful to use a neutral term, as we have not been able to make huge progress in knowledge about aetiology, therapy and other facts concerning this syndrome within 50 years. When Dr. Landau was asked which term he would prefer he said that you should not waste time thinking about terminology. For himself he would not care about the label.

History of the syndrome

The neurologist Landau and the educationalist Kleffner described in an interdisciplinary work, which was published in 1957, the disorder by describing six cases. Frank Kleffner was the head of the department for aphasic children at the "Central Institute for Deaf Children" in St. Louis. One of his colleagues was also Mildred McGinnis, who is known to Speech and Language Therapists because of her work about the association method.

Landau reported that Dr. Silverman, director of the "Central Institute for Deaf Children" was very interested in research work. Together they considered doing a neurophysiologic analysis and so examined 150 patients by using EEGs. During data analysis they identified a small group of children who were different from other children. Landau emphasized that it was Mildred McGinnis who recognize these children.

Landau refers to this story in his article of 1991 "The conception and embarrassing birth of an eponym". However, it was indeed news to me that McGinnes was the one who pointed out to a group of children with loss of speech and epilepsy.

Definition

Based on the classic description of 1957 two main symptoms have always been mentioned when discussing about the LKS:

1. acquired aphasia
2. changes in EEG

Landau answered the question about **his** definition short and undoubtedly, I quote “Children are the ones losing their ability to speak and suffer from insults. That is what we have seen.” In addition the six original cases define the following obligatory symptoms:

- Except from aphasia and epilepsy no further neurologic symptoms become evident
- Landau and Kleffner report that intellectual capacity was not affected in the cases they examined
- In most cases people report about visible epileptic seizures
- Many children additionally have behavioural disorders.

It has not been clarified, whether the level of intelligence has to be maintained, like often emphasised in literature on the basis of the first description.

50 years after the publication of the original paper Landau’s opinion is that the intelligence of the children would be limited.

A narrow definition of LKS should only include children with an original regression of speech. The associated behavioural disorders as well as cognitive impairments are secondary effects of language disorders and can neither be explained as original regression of behaviour nor as regression of the cognitive level.

Are behavioural disorders to be interpreted as secondary effect of language disorders?

Landau emphasizes that these children very often show behaviour disorders. He is not sure though if they are a result of the language disorder.

LKS occurs in early childhood. The exact beginning has been described differently and ranges from the 18th month of life to the age of 13. The age of main manifestation of this aphasia ranges from the age of 3 to 8. The syndrome never manifests in adults but they could suffer from language problems due to LKS. Prognosis of recovery is not always possible, as assumed by Landau & Kleffner in 1957. Boys are affected twice as often as girls.

Aphasic symptoms

At the beginning, the most frequently noticeable symptom is that the ability to comprehend speech decreases. After this, the expressive language also starts to vanish. Isabelle Rapin and her team, like some other authors think that this language disorder should rather be seen as an auditory agnosia, which can appear in different grades of abnormal language production than as a real aphasia.

Children are unable to recognize and decode verbal material. If it is only an agnostic disorder which is fixed on verbal material, you can speak about a verbal auditory agnosia or “word deafness”. This is the reason why the syndrome is also called “acquired auditory verbal agnosia”.

In reference to the question whether it is a real aphasia – a linguistic disorder – or an auditory agnosia Landau asks how to distinguish an auditory agnosia from a linguistic aphasia. He makes aware that “instead of worrying if its aphasia or agnosia, it should describe acquired loss of language.”

A homogeneous pathology of language production disorders concerning LKS cannot be found in any description of a case in literature.

While some authors describe fluent aphasia with paraphasia and neologism, others think language disorders are part of non-fluent aphasia.

One can read frequently that it is a special form of a childhood aphasia, because speech production is fluent with lots of semantic and phonematic paraphasia, neologisms and jargon. However, the opinion that aphasia in childhood are only non-fluent is out-of-date. Every symptom in aphasia which affects adults can also be found in children – fluent forms of aphasia, too.

The beginning of regression of speech in children can be very variable. Landau and Kleffner report in their six cases that aphasic symptoms occurred within days to months. Due to this fact aphasia can start suddenly or develop gradually. Aphasia typically shows a fluctuating development. The ups and downs concern severity and variability. However, there are also static developments. Comprehension problems of speech are typical for LKS.

In reference to the question whether the cases which in literature only show expressive disorders are atypical cases for LKS, Mr. Landau says, I quote "I don't know. Because nobody done proper studies."

So Landau doesn't answer the question whether cases with only expressive language disorders can also be LKS.

Therapy

Three forms of therapy are described:

1. medical treatment
2. speech language therapy
3. surgical therapy

I wanted to know if Professor Landau was in favour of surgical intervention. Professor Landau reacted fiercely to this question He emphasized that he considers brain surgery on children with diseases we do not understand as malpractice and unethical.

In his commentary which was published in "Neurology" 2004 Landau stressed that when there is an unspecific aetiology and a very variable prognosis "uncontrolled, destroying surgical adventures cannot be accepted."

My main question was if LKS should be defined as a disorder with a broad spectrum of symptoms or as a rather heterogeneous or homogenous syndrome?

Landau's answer was clear and he took position for an open definition of LKS.

Specific research is necessary. Dr. Landau's response to my question what he thought of an international data base to classify cases was that we "need is a research program, not a database... a database will do nothing unless its highly systematic, organized... it would be very expensive to do it well."

In a commentary published in 2004 Landau emphasised the necessity of a multi centre study. Strictly defined subgroups would make it possible to conduct studies about aetiology, no matter if they are viral, toxic, genetic or something else. With a sufficient amount of patients it would be possible to conduct strict studies with respective control groups (Landau 1992). 50 years after the first description of the Landau-Kleffner syndrome many questions are still open even for Dr. Landau.

Literature

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