Sign language in Landau-Kleffner syndrome

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SUMMARY

This article reviews the history of sign language (SL) and the rationale for its use in children with profound auditory agnosia due to Landau-Kleffner syndrome (LKS), illustrated by studies of children and adults followed for many years and rare cases from the literature. The reasons that SL was successful and brought some children out of isolation while it could not be implemented in others are discussed. The nowadays earlier recognition and treatment of LKS and better awareness of the crucial need to maintain communication have certainly improved the outcome of affected children. Alternatives to oral language, even for less severe cases, are increasingly accepted. SL can be learned at different ages with a clear benefit, but the ambivalence of the patients and their families with the world and culture of the deaf may sometimes explain its refusal or limited acceptance. There are no data to support the fear that SL learning may delay or prevent oral language recovery in children with LKS. On the contrary, SL may even facilitate this recovery by stimulating functionally connected core language networks and by helping speech therapy and auditory training.

KEY WORDS: Landau-Kleffner syndrome, Acquired epileptic aphasia, Auditory agnosia, Sign language, Manually coded language.

Children with verbal auditory agnosia are deaf in their brains, not in their ears

Isabelle Rapin

The review of sign language (SL) use in Landau-Kleffner syndrome (LKS) (Landau & Kleffner, 1957) constitutes a fascinating and often tragic journey in the history of ideas about the nature, cause, and outcome of this disorder, as well as into the persistent professional controversies about oral versus sign language education for the deaf in the second part of the twentieth century. Children with profound verbal auditory agnosia (VAA) due to LKS are in a similar situation as the profoundly deaf. No doubt all large institutions for deaf children have always taken in one or several of these children, and this became apparent when the condition was recognized as a specific syndrome (Landau & Kleffner, 1957). Schools for the deaf were in the majority oralist in their approach, and the use of SL was forbidden (Lane, 1993). Actually, Frank R. Kleffner, coauthor of their initial paper and former Director of the Central Institute for the Deaf in Saint-Louis, Missouri, U.S.A., where the patients came from, did not mention SL in the case reports or the article discussion.

In her seminal article on VAA in children, Isabelle Rapin—the world’s child neurologist with the greatest expertise in childhood deafness, and an uniring advocate for the Deaf community and of the use of SL (see preface of the book Seeing Voices, Sacks, 1991)—described in detail four children who actually suffered from LKS (this eponym was not yet used at the time) (Rapin et al., 1977). One of them was learning SL, the abstract states: “One child illustrated the close association between writing and phonologic encoding and decoding operations, and two children the preservation of linguistic skills, provided the acoustic channel was by-passed and language presented visually. This latter point has been emphasized because of its implications for the remedial education of children with this syndrome” (Rapin et al., 1977). Since then, the help of some sort of manually coded language was regularly mentioned as part of LKS.
management in severe cases, but without any detailed or systematic study presented. Several clinicians all over the world who recognized the existence of the syndrome and who took an active part in the rehabilitation of these children, had a success or a failure story to tell about the use of SL. However, literature on this issue has remained scarce despite the increasing recognition of and interest in this condition (Worster-Drought, 1971; Cooper & Ferry, 1978), and systematic group studies, let alone controlled studies are lacking.

Actually, studies on LKS came mainly from two types of professionals: on the one hand, child neurologists and “epileptologists” concerned with the medical aspects of the disorder, on the other hand, speech and hearing specialists and neuropsychologists whose main focus was on language (Dugas et al., 1982). The latter reported often extensive studies on oral and, more rarely, on written language difficulties and their evolution over several years. Few were interested or had any significant data to present about SL in their patients, although its usefulness was mentioned. At the time education for the deaf was predominantly oralist, the necessity of a manually coded language was not much advocated. Even now that its use has been recognized as important for children with severe developmental language disabilities, it is not a universal part of the training program of speech therapists.

The silence on the issue of SL in LKS may not reflect the reality of its use and the benefit that some children received. No doubt the difficulty to assess SL skills also prevented the publication of data in scientific journals. The rare studies came from neuropsychologists primarily concerned with theoretical issues, such as language development in the case of auditory input deprivation during a critical period of development (Bishop, 1982; Baynes et al., 1998).

In a follow-up study of “receptive aphasic ex-pupils” of Moor House School—a residential school for children with severe disorders of speech and language in England (Ripley & Lea, 1984)—that included nine adults, born between 1945 and 1953, with “acquired receptive aphasia,” (based on the description, most probably LKS cases) the following conclusion was made about communication: “Without exception, the receptive aphasic ex-pupils were keen to communicate. Speech skills were pushed to their limits; signing and finger spelling were used when speech failed.” Most of young adults had been introduced to local clubs for the deaf, but few had chosen to stay (2 of 9). In the last few years, British Sign Language (BSL) has been introduced at Moor House School as part of the curriculum for older receptive aphasic children. It would be interesting to see if, consequently, more receptive aphasic ex-pupils would become long-term members of clubs for the deaf.

Sieratzki et al. (2001) described a 26-year-old man who lost language at 5 years of age, and who recovered minimal useful oral speech and no lip-reading abilities. From 7 to 12 years of age he was taught in spoken English accompanied by the Paget-Gorman sign system in a school for speech and language disordered children. Only when aged 13 did he enter a school for the deaf where he learned BSL. The latter became his most efficient communication modality with good vocabulary yet severe grammatical limitations. His mother also learned SL and subsequently became a professional interpreter.

In 1989, we reported the adult follow-up study of seven children with LKS (Deonna et al., 1977, 1989), four of whom exhibited severe VAA. They had not received any substitutive language during childhood, they had not significantly recovered, and as adults they remained tragically isolated without effective communication of any kind. This outcome study convinced us even more that introducing a visual language should be proposed despite the many difficulties and the resistance, considering the risk of permanent deprivation of communication in case of absent or partial recovery—which outcome is unpredictable in the early stage of the syndrome.

In 2001, we published a detailed study on SL use in a 13-year-old boy with early and severe LKS, who was educated with deaf children and who later on recovered good oral language (Roulet Perez et al., 2001)—see below, case D.R. with a follow-up at age 25.

Visual languages, SL, and the brain

Sign language acquisition

A child can acquire SL as early and as easily as an oral language. SL, like any oral language, can also be learned as a second language at all ages, with the same variability in the ease of acquisition, even with the presence of increasing difficulties with advancing age. Despite the ancient and persistent prejudice among the hearing community that SL learning may delay or inhibit oral language acquisition and mastery, there are no data to support this claim (Gordon, 2004). On the contrary, several observations including some in LKS children suggest the opposite (Deonna et al., 1989).

Manually coded languages (Table 1)

One must distinguish the natural SL used by a deaf community in a given region or country, and the different

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<td><strong>Sign language</strong>: complex visuospatial system used by the deaf community, with its own morphologic (hand configuration, location, and movement) and grammatical rules (space, timing, mimics).</td>
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<td><strong>Signed spoken languages</strong>: manually coded spoken languages (Makaton: based on British Sign Language + symbols; Paget-Gorman system based on English grammar).</td>
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<td><strong>Dactylology</strong>: Manual alphabet.</td>
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<td><strong>Gued-speech</strong>: Aid to phonemic discrimination.</td>
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manually created coded systems with the same grammatical structure as oral language, such as signed French, signed English, or the Paget-Gorman sign system. SL has its own linguistic structure at phonologic, morphologic, and syntactic levels, derived from different components like hand shapes, hand orientation, location, and movements in function of time and space (Poizner et al., 1987). In children with LKS who were offered a manually coded language, most used a signed system based on oral language, whereas SL was used mainly by those integrated with the deaf. Some children used both systems, simultaneously or consecutively. This has been rarely detailed in the papers dealing with nonverbal communication in these children.

**SL and the brain**

Congenitally deaf signers who sustained focal brain damage have shown the same left-sided hemispheric dominance for SL. More recently, functional imaging studies comparing hearing monolinguals, deaf signers, and bilingual hearing children of deaf adults in different experimental conditions have shown common patterns of activation, involving the dominant frontal and temporoparietal regions. (Sakai et al., 2005 for review) concluded that “these results demonstrate amodal commonality in the functional dominance of the left cortical regions for comprehension of sentences, as well as the essential role of the left F3t/F30 (ventral part of the inferior frontal gyrus) in processing linguistic information from both signed and spoken sentences.” Brain activation patterns of hearing people presented with speech and deaf people presented with SL showed of course some differences that reflected the modality of input and not the linguistic aspect of the task (Campbell et al., 2008).

**LKS: Rationale for SL or other visually coded language use**

Children with LKS initially have an auditory agnosia which, in severe cases, means that no sounds can be recognized. When they improve, the decoding of environmental, long-standing auditory agnosia, the child who is proficient in SL or in another manually coded language may not be interested in sounds or may have “forgotten” that speech sounds can have a meaning. Once recognition of sounds and simple words is again possible, a systematic auditory training as proposed by Vance and colleagues (Vance, 1991, 2001) can be started, while maintaining the use of SL and explaining to the child the reasons for this undertaking.

### Outcome of Children with Severe Verbal Auditory Agnosia: Personal Observations and Case Illustrations

**Historical adult case illustration (Deonna et al., 1989)**


RG lost all comprehension and expression of language when aged 3½, and has never recovered since then. As a child, a hearing aid was tried. She remembers it as a negative experience. She never had contacts with the deaf community. The family recalls having visited a school for the deaf with her, but she refused to enter it and this was never discussed again. She attended a special school for mentally deficient children with whom she had no contacts. The diagnosis of LKS was made when she was 10 years old. At that time she had a WISC Performance IQ of 87. At age 12, she started learning the French SL basics with her speech therapist, who learned them together with her. At age 20, she joined an SL course organized by the deaf community. She enjoyed it very much and made significant progress. A video taped discussion with her mother was shown at that time to a bilingual SL teacher who noted that she used some elaborated sentences and that the placing of signs was at a high level for a beginner. He thought that she was an intelligent person who would certainly progress in SL. Since the age of 20 years, she has been employed as a laundress at our hospital, where her senior colleague is a profoundly deaf person with whom she communicates in SL. She otherwise never had regular contacts with other deaf people. She married a hearing man, got a driver’s license, and is fully independent. She has a 13-year-old son with whom she communicates using SL. She has a rudimentary knowledge of written language,
understands simple words and questions, and uses short
text message. In 2007 she was seen in a café chatting
quickly in signs with a young man. Her mother thinks
she is far behind in SL as compared with deaf people.

C.S., born 1964, 45 years old. Severe permanent
auditory agnosia. No sign language communication
despite years of schooling with the deaf (case 5, Deonna
et al., 1989).

CS had onset of VAA at 4 years. From the age of 5 to
17 years, he was in two different schools for the deaf and
lived with the profoundly deaf but did not learn any sig-
nificant SL. At 42 years of age, he lives in a sheltered
home and communicates with a few simple words he has
retained and a very basic understanding of common
words almost exclusively with his family. His parents
were against SL from the beginning. He never wanted to
be with the deaf and to use and learn signs. He reads few
words (newspaper headlines). He understands and knows
few elementary signs. This almost total absence of ges-
tural communication despite his years of living with the
def is hard to understand. In the seventies, the approach
was purely oralist and he did not identify with the pro-
foundly deaf who signed, did not consider himself (nei-
ther did they) part of their world. The deaf pupils who
were fluent signers may have excluded him, especially if
he was not skillful or slow in learning. He is certainly not
retarded or autistic, but tended to withdraw and be
aggressive as a child. It appears unlikely that he had spe-
cific difficulties with using gestures as language symbols,
although this ability was never really put to the test. One
can only suppose that this tragic outcome was the com-
bined result of parental resistance, ambivalence of the
teaching staff who insisted on the oralist approach, and
absence of identification and possible rejection by the
profoundly deaf. His always preserved, albeit very rudи-
mentary, oral comprehension and expression and the
expectation of progressive improvement probably also
gave a false hope.

More recent adolescent and young adult case

D.R., born 1983, 25 years old. Total auditory agnosia
with recovery of oral language. Fluent SL, educated with
the deaf. Bilingual (Roulet Perez et al., 2001)

This boy lost all language when aged 2½, but the diag-
nosis of LKS was made only at age 5½. Anticipating what
his future might be, and thanks to the efforts of several
professionals and the final acceptance of the family
despite early reluctance, he was admitted to a school for
the deaf at age 6. There he spent his entire school years.
When aged 13, a study of his SL skills compared to those
of a congenitally deaf child was performed (Roulet Perez
et al., 2001), and showed a remarkable mastery of SL. In
addition, auditory training as advocated by Vance and
colleagues (Vance, 1991, 2001) was undertaken from the
age of 9 years on. His motivation and cooperation in this
arduous work were initially low but increased gradually
when the aim of this training was repeatedly explained to
him using SL. He experienced a very good recovery of
oral language, which allowed him to receive a further
secondary school education and vocational training as a
designer in a technical school. He also learned a fair
amount of English and German, and exhibited functional
written language skills despite far from normal speech
decoding and production abilities. He has kept friendships
in the deaf community.

This study, albeit a single case, showed that the ability
to develop a linguistic code in SL can be fully preserved
in LKS. Although this may not be possible in all children
with LKS whose epileptic dysfunction may extend beyond
the central auditory cortex, this case clearly shows that
another form of language can be totally mastered, a possi-
bility that has been often questioned. Even more impor-
tantly, SL did not prevent the recovery of oral language
but possibly even facilitated it, as soon as the remission of
the epileptic disease made it possible. The patient’s life
story has been told on television in a regular series for the
def ( Télévision Suisse Romande, 2006).

C.L., born 1996, 12 years old. Rapid learning and success-
ful transient use of SL with full oral language recovery

This boy was 4 years old when he lost all language
comprehension and expression within a few months. He
soon started to use natural signs. About one year after dis-
 ease onset, formal SL (a combination of French SL and
signed French) was introduced. His mother, his 9-year-old
sister, and the speech therapist learned SL as well, with a
surprising enthusiasm. He was kept in his regular elemen-
tary school. He rapidly became proficient in SL, his sister
even more so (Télévision Suisse Romande, 2006). With
steroid treatment, significant verbal recovery occurred
about 6 months after introduction of SL (see figure),
resulting in a spontaneous decrease of its use. SL had been
his main means of communication for about one year; it
was subsequently gradually less necessary for another
year. He fully recovered oral language and is off all treat-
ment since 10 years of age, with a normal sleep and wak-
ing EEG (Dr. Pierre Genton, Marseille). His family is
convinced that SL was a crucial factor in keeping him a
full member of the family, allowing the expression of
feelings and the ability to refer to other subjects than the
elementary “here and now.” At age 12 his IQ is in the
upper norms (Perpetual Reasoning Index [PRI] = 107,
Verbal Comprehension Index [VCI] = 112). Formal neu-
ropsychological testing showed normal written and arith-
metic skills. The only deficits affect the short-term verbal
memory span and high level phonologic skills. Dichotic
listening tasks show total right ear extinction, as was the
case previously.
Prolonged communication deprivation during childhood

M.I., born 1986, 19 years old. No manually coded visual language use. Good late oral language recovery

This girl lost all verbal comprehension within a 2-week period at 3.2 years of age. The diagnosis of LKS was rapidly made, and she was treated with steroids for several years with EEG improvement. However, she continued to display severe comprehension difficulties. It was only after 9 years of age that she became able to have a small conversation. A manually coded language was proposed early, but was never implemented. She followed school in her village where she had no friends and felt rejected. Her parents thought that her condition was not understood. We followed her regularly with formal language and cognitive tests from age 11–19, and had contact with her parents, speech therapists, and child psychiatrist. She has made a remarkable partial recovery of oral language with intensive training. She currently works as a kindergarten assistant and lives independently. Only at the age of 19 did she have enough verbal language and insight into her own disability to express her desire to know all about her disease. The analogy with deafness and the fact that some children used SL with success, however, were a source of irritation and could not constitute a topic of discussion for her or her parents. M.I. and her parents were quite angry with the school where she had been totally isolated. She never had close friends until recently, and does not want to associate with her former classmates.

**DISCUSSION**

The idea that children with LKS, who once did possess a normal oral language and were not deaf should rapidly learn SL or a signed oral language, is difficult to accept and even more so to implement. It also requires the commitment of the whole family and professionals. To feel associated with the deaf community is also sometimes strongly rejected. When there is some residual speech or when language is fluctuating, this is even more difficult to accept. In these circumstances one should not be discouraged but on the contrary, one should keep in mind how successful SL was in some children and their families. SL allowed avoiding the tragic cutting of communication, and maintained open channels for sharing information, expression of emotions, and school learning. It also became an opening into another world and another culture, and was a source of personal pride.

We have seen very variable parental responses to the suggestion of introducing SL: from immediate and definitive rejection to initial reluctance with progressive acceptance as well as to enthusiastic and natural understanding and willingness to do whatever could help maintain contact with the child. In the days following the diagnosis in her 6-year-old child with recent onset of auditory agnosia, a mother wrote in her diary at the time antiepileptic treatment had just been started: “August 4: while putting him to bed, I speak to him about sign language. I have the impression that he would like it, only the fact that this would be far from home worries him.” In his case, language comprehension returned quickly under prednisone and he never had a relapse thereafter.

The drastic changes in behavior, mood, well-being, and family relationships observed in these children when communication became possible through a means they could master—sometimes after a long time of deprivation—has perhaps been the most important lesson. Several examples show that SL can successfully be learned at different ages with a clear benefit. Finally, SL did not interfere at all with oral language recovery, as shown in several children. SL may even have facilitated recovery by stimulating functionally connected core language networks. In addition, SL allows parents and professionals to explain to the child who is recovering some oral comprehension, why auditory training is that important. In doing so, the child does not remain a mere passive recipient of exercises he cannot understand the use of.

The most difficult aspect of SL learning concerns the ambivalence that most children and their families experience in their relationship with the world and culture of the deaf. This mirrors the educational problems of deaf children born from hearing parents, and the still-existing prejudices against SL. The sometimes preserved and fluctuating oral language abilities in conjunction with the efforts made in speech therapy, as well as the fear that SL would delay or prevent oral language recovery, probably all contribute to the limited acceptance or even the refusal of SL—as well as to its often poor mastery.

Language is the main defining feature of a culture. Even in the best circumstances it can be hard to belong to, to feel at ease with, and to develop a strong identity feeling while having to live in different cultures. This is the issue of biculturalism. It is difficult enough to master two oral languages when one is perceived as an inferior one and is not positively experienced by the learner. Infrequent practice and lack of motivation can of course lead to a poor efficiency and an unsatisfactory feeling. This applies to many situations of bilingualism (Grosjean, 1996). Case D.R. is rather an exception in this respect (Roulet Perez et al., 2001). The practical feasibility of SL learning can be another hurdle, as neighboring schools for deaf children, close to the child’s home, are often not available. Integration in a regular school with the help of a signing speech therapist—which is a skill that some of the younger generation are keen to learn—may be an alternative option. Of course, it is not possible to state at what age or severity degree of auditory agnosia a manually coded language should be introduced—or is it possible to predict its type: SL or just signed oral language. Only individual decisions

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can be made. Children with LKS sometimes exhibit severe behavioral problems and fine motor/praxic difficulties, which may hamper SL learning. Some may have specific linguistic difficulties in SL, but this is, of course, not predictable (Metz-Lutz et al., 1999).

In future studies, one should make efforts to monitor progress in SL, using for instance a systematic protocol and serial videotaped recordings of the child’s comprehension and production of SL (as done with oral language), with the help of competent signers (see Roulet Perez et al., 2001) in order to document the learning of new signs, rapidity of acquisition, and analyze types of errors.

**Conclusions and Broader Implications**

Most probably the unfortunate fate of some of the patients described in this review will no longer exist thanks to the earlier recognition of LKS nowadays, the active and prolonged medical treatment, the increased awareness of the crucial need to maintain communication by all means—including at some point the exclusive use of a manually coded language—and good interdisciplinary work. From this perspective, this article may already represent a piece of medical history, but we thought that the history of these children in the early years before recognition of the disorder had to be put on record; otherwise it might have been lost forever.

On the other side, severe refractory cases of LKS will always exist, and these children will need an alternative to oral language. There are few other examples in human history that are comparable to what is seen in children with severe LKS, who have totally lost their previously normal ability to communicate orally, have learned a different native language, thus becoming bilingual. Important issues like successful delayed restoration of unused cortical functions or the occasional occurrence of good reading without phonology or the reorganization of brain functions after deprivation, which are difficult to approach empirically, could also be studied in patients with severe LKS who use SL.

**Acknowledgments**

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**References**


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