**Introduction**

LS is an enigmatic epileptic condition, characterised by loss of control over behaviour, speech and involuntary movements. It usually begins in children aged 3 to 7 years with symptoms of auditory hallucinations, leading to severe communication and comprehension disorders. Impeccate and primary hearing components are usually preserved in the early phase of the disease. Within a few months his speech language regressed to the level of a 2 year old child, and has not recovered since. His childhood EEG showed severe epileptic aberrations over the hemisphere, with some spread to the left.

When age 12 SC attended a school for children with speech and language disorders and was taught to speak English accompanied by the Paquet-Garman Sign System. From age 12 - 20 he attended a residential school for the deaf where he was introduced to ISL. He worked as a gardener and lived independently.

As an adult, SC recognises a small set of familiar words and short phrases through his gross sound contour, without phonological decoding. He cannot lip-read. SC’s speech is generally unintelligible except to those who know him well. His reading is too slow for efficient communication. He recognises graphic pattern of whole words “by eye”, without awareness of grapheme-phoneme correspondences. SC can identify familiar voices, as well as many environmental non-speech sounds. He enjoys music and recognises melodies, but has difficulties in retaining and reproducing rhythms. He communicates well in ISL with normal vocabulary but with specific limitations in ISL grammar, typical of late learners of sign language as a primary language end of signs with dominant hemisphere damage.

SC has a surprising knowledge of English phonemes. During a test, he responded to the target picture with correct English, then explained in ISL that the word “watch” could also mean “to see”.

Previous functional imaging studies in adult patients with LS have shown mismatches in language activations, supporting the concept of functional ablation originally proposed by Landau and Klerffer. However, in these studies, patients were tested, and not challenged by any language processing tasks.

We have now conducted two comprehensive linguistic and MRI case studies of patient LS, with which our clinical activity was measured during language tasks.

The most prominent findings are:

- A significant advantage of British Sign Language (BSL) over spoken and written English very strong temporo-parietal activation in response to spoken English. Suggesting a high level of activation potential rather than a state of functional ablation in a distinct pattern of processing of BSL.

The pattern of MRI findings in combination with comprehensive language performance tests have provided unique insights into the neuroanatomy of the pathologies of ISL and further justification for the use of a sign language of the deaf in patients affected by this condition.

**CASE STUDIES**

**Language assessment**

<table>
<thead>
<tr>
<th>Age SC</th>
<th>Standardised Age Equivalent</th>
</tr>
</thead>
<tbody>
<tr>
<td>5'6&quot;, 13.0&quot;, 2'1&quot;</td>
<td>2.0, 2.2, 2.4</td>
</tr>
<tr>
<td>15'-0&quot;</td>
<td>0.0</td>
</tr>
<tr>
<td>6'7&quot;, 10'-0&quot;</td>
<td>&lt;5.0, 7.0</td>
</tr>
<tr>
<td>11'-0&quot;</td>
<td>&lt;5.0, 7.0</td>
</tr>
<tr>
<td>11'-0&quot;</td>
<td>&lt;5.0, 7.0</td>
</tr>
</tbody>
</table>

| English comprehension | 9'-7", 10'-1", 10'-8" |
| English production | 7'-6", 9.0 |
| Word Reading | 6'7", 10'-0" |
| Word spelling | 6'7", 10'-1", <5.0, 7.0 |
| BSL Vocabulary | 11'-0" |
| BSL Grammar | 11'-0" |


**Discussion**

Speech and language therapy

Children with severe LS should have the opportunity to learn a natural sign language of the deaf. This will not only improve their communication skills, but also improve their self-esteem. It can be seen that ISL has a higher communicative capacity than other artificial sign languages and does not depend on the disturbed speech systems.

ISL vs. speech and visual forms of sign language

Access for any form of spoken language to the primary auditory system is required. However, sign language may provide an alternative pathway. This reflects the natural advantage of ISL over the auditory pathways, in terms of the auditory systems of spoken language.

How is the speech crisis affected in ISL?

Landau and Klerffer originally proposed a “functional abolition” of the speech crisis, as a sequel of the severe epilepsy. This view is not supported by our study, which shows strong activation in these children in response to speech input.

As first suggested by Morrell, epilepsy may interfere with the normal process of synaptic pruning, and thus create an excess rather than a reduction of synaptic connections. The impairment in ISL is seen as the product of abnormal, developmental remodelling in the speech crisis. This explanation is not in agreement with the observation of rapid restoration of previously acquired speech skills, after neuro-surgical treatment.

We believe that the impairment in ISL originates in a functional disturbance of the speech crisis. This is manifested in hyperactive responses to the demands of speech processing, triggering epilepsy, inserting with synaptic pruning, enhancing formation of recurrent excitatory cortical loops, and automatic, normal remodelling of the developing brain. This state may exist without clinical epilepsy and persist after puberty.

**Literature**


**References**


**Viewing BSL signs:** Activation predominantly on the right (RH) extending from occipital temporal cortex (Brodman area (BA) 37), posteriorly into the middle occipital gyrus (BA 19). Superiorly into the medial occipital gyrus (BA 18).

**Hearing words:** Extensive and powerful bilateral, though stronger right sided, activation of primary (BA 41) and secondary (BA 42 & 22) auditory cortices. Additional activation in the intraparietal gyrus (BA 40) and upper aspect of Broca’s homologue in the RH (BA 44/45). The extent and power of activity in the auditory cortex far exceeds that when hearing music (not shown here).

**Viewing BSL signs:** Activation exclusively in left posterior temporal regions (BA 37 & 39), with very limited activation in MTG (middle temporal gyrus).

**Hearing words:** Extensive and powerful activation was observed in left posterior perisylvian areas (BA 22, 39/40) with the apparent exception of BA 42/44 in contrast there was consistent right hemisphere activity limited to BA 42/44 (Heschl’s gyrus).

The extent and power of the activity far exceeds the activation in other language tasks: BSL viewing written words, lip-reading words (not shown here).