Review

The neurology of sign language

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Abstract

Forms of sign language have developed in a number of countries. American Sign Language, which originated from French signing, has been most extensively researched. As sign language is based on gestures executed in space and perceived visually it might be thought that it would mainly be a function of the right cerebral hemisphere when this is the non-dominant one. A number of studies are reviewed showing that sign language is a language in its own right and therefore, as with spoken language, its primary site of organization is in the dominant hemisphere. This does not mean that there is not a significant contribution from the other hemisphere with an interplay between the two. Each research project usually contributes some facet of knowledge apart from the main conclusions. These included the importance of distinguishing signs from gestures, the localization of different types of signing within the left dominant cerebral hemisphere, the fact that lesions of the right non-dominant hemisphere, although not causing a loss of signing will result in dyspraxia, and that aphasic symptoms of signing and speech are not modality dependant but reflected a disruption of language processes common to all languages. Examples are given of discoveries made by the use of the newer neuroradiological techniques such as functional magnetic resonance imaging and positron emission tomography, and no doubt these will lead to further advances in knowledge. The use of sign language in the treatment of patients with verbal aphasia is considered, especially of children with the Landau–Kleffner syndrome, but therapy of this kind can be used in children with delayed language development, and in other types of acquired aphasia at any age. Other methods of treatment than signing, such as cochlear implants may be increasingly used in the future, but it seems likely that sign language will continue to be a dominant feature in the deaf culture.

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1. Introduction

The history of the development of sign language goes back for more than 400 years, so it is not surprising that a complicated culture of the deaf has arisen [1]. What is surprising is that until a few years ago there were bitter arguments between those who favoured oral language only when communicating with those who were severely deaf and the supporters of signing. Some of the former would not allow the use of sign language, which never seemed to be very logical, as surely an input of language, whatever form it took, should be of benefit. A form of sign language has developed in at least 13 European countries, but some of these are mutually unintelligible and may have little or no similarities with the local spoken language. Most research has been done on American Sign Language, which originated from French signing about two hundred years ago, and this has shown that it is indeed a fully-fledged language that can be used in every day conversation, intellectual argument, wit and poetry [2]. In the case of wit punning is a good example. This has been studied among subjects using American Sign Language, and depends on the similarity between signs which have different meanings. With poetry the subjects were those associated with the National Theater of the Deaf who used the same signing language, and was based on an analysis of the flow of movement between signs and the patterns of movement [3].

Also, the study of the neurology of signing can help to discover the mysteries of language in general [4]. For instance the work of Petitto and Marentette [5], who showed that deaf infants of deaf parents whose only language exposure was to American Sign Language engage in a form of babbling which is quite like the babbling of hearing infants. Around about the age of one the former produced manual activities different from the typical gesturing of
normal children at this age, showing that babbling is most probably related to the abstract linguistic structure of language rather than the speech modality per se.

2. Neuronal basis for sign language

In most instances there is no doubt that the left cerebral hemisphere dominates the organization of spoken language, and that the right hemisphere is mainly concerned with visual-spatial functions. Therefore as sign language is based on gestures executed in space and perceived visually it might have been thought that it would be mainly a function of the right side, but this does not seem to be the case. Also in studying such functions sign language must be clearly distinguished from gesture. Metz-Lutz et al. [6] reported a congenitally deaf child, aged five, with focal epilepsy. He showed specific impairment in French sign language but an amazing performance in miming, which supports an early dissociation of the two systems, verbal and non-verbal. This does not mean that the right cerebral hemisphere does not have a part to play in the functional organization of sign language, just as it does with spoken speech, for example in automatic speech such as repetition. Neuroimaging studies have shown that in the case of sign language the right cerebral hemisphere is involved in such areas as discourse comprehension, prosodic functions, and in the direct encoding of objects [7]. Also, in deaf signers damage to the right hemisphere may not result in aphasia but will cause gross impairment of non-linguistic visual-spatial abilities, and as with hearing subjects left hemisphere damage can cause subtle difficulties of this kind in addition to the linguistic effects [8]. As stated by Maratsos and Matheny [9] language function is most unlikely to be a wholly independent one.

Most right handed people are left hemisphere dominant for language; and Vaid et al. [10] tested the performance of the right and left hands of deaf native users of American sign language. They found that more signs were produced with the preferred hand when right and left handers were tested, but left handers were more flexible in signing with their non-preferred hand and more unusual patterns of hand use were found in a right-handed deaf signer after a left hemisphere lesion. This suggests an increased use of the intact right hemisphere. Also it has been found that although much of sign language function is organized in the left cerebral hemisphere, even within these areas the exact localization of any lesion will result in different types of dysphasia, for example whether this affects vocabulary or grammar [11].

Damasio et al. [12] studied a hearing, strongly right-handed, woman who had learnt American Sign Language at the age of eighteen. The left intracarotid injection of a barbiturate, the Wada test, was used, before and after a right temporal lobectomy for intractable epilepsy. Neuropsychological and anatomical asymmetries favoured left cerebral dominance for auditory based language. Single photon emission tomography showed lateralized activity of the left Broca’s and Wernicke’s areas for spoken language. During the Wada test these areas would be inactivated, and the patient was rendered aphasic for both English and sign language, both before and after the operation. The results therefore suggest that structures in the left cerebral hemisphere subserve ‘language’ in a visuo-spatial as well as an auditory system, rather than individually on speech sounds or the form of the signal.

Similar findings were recorded in a hearing 19-year-old raised by deaf parents who suffered a head injury. There was severe right hemiplegia. He had no evidence of apraxia or visual-spatial deficits, but was aphasic for the spoken word and for sign language [13]. In contrast an adult patient, after a right cerebral vascular lesion had no disturbance of sign language or finger spelling [14].

In a study of language processing in various forms MacSweeney et al. [15] used neuroimaging with functional MRI to explore the perception of British Sign Language in nine hearing and nine congenitally deaf native signers while they performed a British Sign Language sentence-acceptability task. Eight hearing, non-signing subjects performed an analogous task that involved audio-visual English sentences. They found that there were modality-independent patterns with regions activated by the sign language among the deaf and by spoken English among the hearing. These included the inferior prefrontal regions bilaterally and the superior temporal regions bilaterally; and lateralization patterns were similar for the two languages. There was no evidence of enhanced right hemisphere recruitment for the sign language processing in comparison with audio-visual English. In relation to modality-specific patterns, audio-visual speech in hearing subject generated greater activation in the primary and secondary cortices than the sign language in the deaf signers, whereas the sign language generated enhanced activation in the posterior occipito-temporal regions, reflecting the greater movement component of sign language. Then the influence of hearing status on the recruitment of sign language processing systems was investigated by comparing deaf and hearing adults who had British Sign Language as their first language, and the former showed greater activation in the left superior temporal gyrus in response to this language (native signers); suggesting that this region may be privileged for processing heard speech even in the hearing native signers, and that in the absence of auditory input this region can be recruited for visual processing. In fact it shows that these regions constitute the core language system regardless of the language modality and hearing status.

Research investigating the neural organization of signed language and spatial cognition in deaf people by Hickok et al. [16] has shown the same hemispheric asymmetries found in those with normal hearing and speech. However, the hemisphere organization of grammatical aspects of language seems to be independent of the modality, and to be unaffected by the significant degree of visual-spatial
processing used in sign language. Evidence from cerebral
lesions among deaf signers suggests that there is a high
degree of similarity in the neural organization of signed
and spoken language within the left cerebral hemisphere, and
that any slight differences that may occur are due to
variations in input–output systems. For example, a
congenitally deaf left-handed man suffered a stroke in
later life, resulting from a left middle cerebral artery
thrombosis. Apart from the right hemiplegia, he lost his
ability to communicate with sign language so that even
though he was strongly left-handed it is reasonable to
suppose his language function was located in the left
cerebral hemisphere [17].

Chiarello et al. [18] investigated a sixty five year-old
prelingually deaf woman who was fluent in American Sign
Language. She had suffered from a left parietal infarct
involving the left supramarginal and angular gyri which
resulted in aphasia for both the sign language and written
and finger-spelled English. By five to seven weeks after this
there was fluent but paraphasic signing, anomia, impaired
comprehension and repetition, alexia and agraphia with
elements of neologistic jargon. In general the patient’s sign
errors showed a consistent disruption in the structure of
American Sign Language which paralleled the speech errors
of oral aphasic patients. It was concluded that most of the
aphasic symptoms were not modality dependant, but
reflected a disruption of linguistic processes common to
all human languages.

Three hearing adults studied by Anderson et al. [19]
suffered from severe aphasia, but became competent in
certain aspects of American Sign Language and finger
spelling, although virtually unable to speak the English
counterparts of the signs. Two had cerebrovascular
accidents and one herpes simplex encephalitis. The former
suffered damage to the left postero-lateral temporal and
inferior parietal cortices, and they mastered the production
and comprehension of simple signs and short meaningful
sequences but the other, with damage to most of the left
temporal cortex, was less accurate in single sign processing
and was unable to produce sequences at all. The findings
suggested that conceptual knowledge is represented inde-
dependently of the auditory vocal records for the correspond-
ing lexical entries. Also that the areas damaged in the third
patient are part of the neural network supporting the links
between conceptual knowledge and linguistic signs, es-
specially as they are used in sequencing. In other words
dysfunction of the primary auditory-vocal system of
communication does not predict dysfunction in alternative
visual-gestural systems.

3. The therapeutic use of sign language

If a child is unable to communicate with spoken speech,
whether the cause is congenital or acquired, the possibility
of granting the opportunity to do this with the use of sign
language must be considered. In a survey of up to 40 schools
and units for autistic and aphasic children in the UK,
published in 1984 [20], it was found that the majority used
some form of augmentative system. The use of the British
Sign Language was favoured for autistic children and the
Faget Gorman Sign System for those with aphasia. The
results were very variable, but were better for the latter.

In the case of delayed language development there seems
to be no reason why the use of sign language should not be
introduced at an early age. Von Tetzchner [21] report a
3-year-old boy who was not talking due to right temporal
agensis. He was taught sign language, and 6 months later
showed a marked improvement in spoken language.

In patients who lose the ability to use spoken speech due
to acquired aphasia, for example in childhood epileptic
aphasia, or the Landau–Kleffner syndrome, sign language
may provide an alternative means of communication. Perez
et al. [22] report such a child. A boy between the ages of 3
and 4 years began to respond less to auditory stimuli, and
deafness was suspected. Then seizures started and this
syndrome was diagnosed, and by the age of 6 the child was
mute. He subsequently developed considerable skills in
using signing, and to evaluate this his performance at the
age of 13 years 6 months was compared with that of a child
with congenital deafness. This showed that he had acquired
the same proficiency as the other child, and that this had not
impeded the return of spoken language, and may have even
enhanced recovery. This must mean that sign language was
processed in spared higher language areas, and supports the
idea that it does not impair the acquisition of spoken speech.

Woll and Sieratzki [23] point out that in using sign
language, depending mainly on spatial features, in the
treatment of a condition such as the Landau–Kleffner
syndrome affected children are not using representations
of spoken language but a quite separate language, which is
likely to be as accessible and natural to them as to severely
deaf children; and this seems to be the case. Sieratzki et al.
[24] maintain, as a result of functional magnetic resonance
imaging studies, that this syndrome is due to a hyper-intense
cortical response to speech input rather than a disconnection
of cortical speech areas due to epileptic activity [25], and
believe that all affected children should be given the
opportunity to learn sign language. If the processing
requirements of language can trigger paroxysmal activity
which blocks the use of spoken language, and the patient
can learn signing, this must surely mean that this form of
communication can be acquired without access to the
primary system in the dominant hemisphere.

In her studies of children with the Landau–Kleffner
syndrome Bishop [26] found that there were strikingly
similar patterns of performance between children who were
profoundly deaf and those with this syndrome, probably due
to both having deviant comprehension because of their
reliance on the visual modality to learn grammar. If, in both
instances, language comprehension is deviant rather than
delayed this must be taken into account in assessing the progress of these children.

Also, as quoted, people with acquired aphasia can be helped to communicate with the use of signs when they cannot use the spoken word [19]. The success of this may depend on the type of language deficit. For example Kirshner and Webb [27] reported an adult who after a bitemporal infarction showed a severe disturbance of auditory comprehension with relative sparing of the comprehension of printed or gestured symbols; and her ability to communicate was greatly aided by instruction in Amerind (American Indian Signs) and American Sign Language. It has been found that the gestures of the former were easier to imitate than the matched signs of the latter. The contribution of the right cerebral hemisphere was small in both instances, and the sign and verbal systems showed evidence of dissociation [28]. These finding have been confirmed by Guilford et al. [29], although they found no differences between the systems. It was not possible to predict the subject’s level of acquisition for sign language by his expressive speech characteristics alone. Also the severer the aphasia the more difficult it will be for the patient to learn sign language [30,31,32], and success depends particularly on three factors; the degree to which a sign’s physical or structural characteristics make its meaning evident, the motor complexity of the signs, and their linguistic function (noun, verb or adjective). This has obvious implications when selecting a sign vocabulary [33].

4. Conclusions

The importance of sign language to the world of the deaf is shown very clearly in Seeing Voices, a fascinating book by Oliver Sacks [34]. The complexity of the deaf culture are described in a way that gives insight to those who have no concept of what it may be like to be deprived of auditory input. This culture must be respected but efforts must still be made to overcome the disability, for example with the use of more sophisticated digital hearing aids, and operative treatment.

There has been much criticism of cochlear implantation from the deaf community, especially in the case of the prelingually deaf children. This is mainly on the grounds either that it does not work, or that there are severe complications, or that it impoverishes language development, or that it is destroying a linguistic minority. However, there is increasing evidence that the assessment of infant’s hearing is becoming more accurate and that the operation is now safer and more successful. Such success is of particular importance when there is improved development of speech reception, language acquisition and reading comprehension, so that such children can take their place in a hearing environment on equal terms [35]. However, the results at this stage are limited. Priesler et al. [36] investigated patterns of communication of 22 deaf children, aged 2–5 years old, who had used implants for between 1 and 3.5 years. The majority of them could take part in simple oral conversations in a well known context, but they did not take an active role in fantasy play with their peers who did not use sign language, and mostly interacted with signing adults. In fact they were still considered to be socially deaf and did not show any improvement in social competence. Obviously, longer follow-up studies are needed as there will be many factors determining future development.

The most important findings in support of signing being a language in its own right are anatomical. A number of these investigations have been reviewed, and two further studies by Poizner and his colleagues [37,38] are of special importance. They assessed four deaf signers, three of whom had damage to the left cerebral hemisphere and one to the right. The patients were tested for dyspraxia and pantomime recognition, and the latter was not apraxic as might have been expected, but all the others were aphasic for sign language and strong dissociations emerged between their capacities for sign language and their nonlinguistic motor skills. Therefore the language deficits of these patients related to the specific language components of sign language rather than to an underlying motor disorder or to an inability to express or comprehend symbols, however it has been argued that there may also be a disordered motor component when it comes to choosing oral musculature for speech or hand muscles for signing [39]. There is surely nothing in this which is incompatible with the ideas on modularity in a discussion by Marshall [40]. In this he states that it is not unreasonable to admit that a module can cut across sensory boundaries.

Studies of congenital and acquired deafness and dysphasia have already contributed to the knowledge of cerebral localization and adaptability, and will continue to do so. Although the results of examinations of patients with brain lesions may be limited it is certain that the use of techniques such as functional magnetic resonance imaging and positron emission tomography will solve many of the problems [41]. For example, Corina and McBurney [42] confirmed that by using cortical stimulation mapping in deaf signers there was evidence that Broca’s area of the dominant hemisphere was specialized for sign production. Also, by means of functional magnetic resonance imaging in deaf signers there was no doubt about the importance of dominant hemisphere language structures, but that the right hemisphere made an important contribution to the processing of signing as well. In fact it is easy to oversimplify hemisphere specialization between language and visual-spatial functions when in sign language there is an interplay between them [43].

References