A CASE OF ‘SIGN BLINDNESS’ FOLLOWING LEFT OCCIPITAL DAMAGE IN A DEAF SIGNER

GREGORY HICKOK,*† EDWARD KLIMA,‡‡ MARK KRICTHEVSKY§ and URSULA BELLUGI†

†Laboratory for Cognitive Neuroscience, The Salk Institute for Biological Studies, 10010 N. Torrey Pines Rd., La Jolla, CA 92037, U.S.A.; †University of California, San Diego, U.S.A.; and §UCSD Medical School and San Diego V.A. Hospital, La Jolla, CA 92093, U.S.A.

(Received 28 December 1993; accepted 1 June 1994)

Abstract—We report on a right-handed, deaf, life-long signer who suffered a left posterior cerebral artery (PCA) stroke. The patient presented with right hemispheric hemianopia, alexia and a severe sign comprehension deficit. Her production of sign language was, however, virtually normal. We suggest that her syndrome can be characterized as a case of ‘sign blindness’, a disconnection of the intact right hemisphere visual areas from intact left hemisphere language areas. This case provides strong evidence that the neural systems supporting sign language processing are predominantly in the left hemisphere, but also suggests that there are some differences in the neural organization of signed vs spoken language within the left hemisphere.

Key Words: language; sign language; spatial cognition; hemispheric asymmetries.

American Sign Language (ASL) exhibits formal structuring at the same levels as spoken languages and has similar kinds of organizational principles as spoken languages. At the core, spoken and signed languages are essentially identical in terms of rule systems. Nevertheless, on the surface, signed and spoken languages differ markedly. The formal grammatical structuring assumed in a visual/manual language is deeply influenced by the modality in which the language is cast, at all structural levels. ASL displays complex linguistic structure, but unlike spoken languages, conveys much of its structure by manipulating spatial relations, making use of spatial contrasts at all linguistic levels [14, 16].

The most striking surface difference between signed and spoken languages is the reliance on spatial contrasts, most evident in the grammar of the language. Instead of relying on linear order for inflectional marking, as in English (act, acting, acted, acts), ASL grammatical processes nest sign stems in spatial patterns of considerable complexity, thereby marking grammatical functions such as number, aspect and person. Grammatically complex forms can be spatially nested, one inside the other, with different nestings producing different meanings. Similarly, the syntactic structure specifying relations of signs to one another in sentences of ASL is also essentially spatially organized. Nominal signs may be associated with abstract positions in a plane of signing space, and direction

*Address for correspondence: Laboratory for Cognitive Neuroscience, The Salk Institute for Biological Studies, 10010 N. Torrey Pines Rd., La Jolla, CA 92037, U.S.A.
of movement of verb signs between such endpoints marks grammatical relations. Pronominal signs directed toward these previously established loci clearly function to refer back to nominals, even with many signs intervening. This spatial organization underlying syntax is a unique property of visual-gestural systems [14, 16].

The present case report is part of a large programme in which we have been examining the neural organization of language through the study of sign language. Our approach involves the study of deaf signers with focal lesions to the left or the right cerebral hemisphere. We investigate several major areas, each focusing on a special property of the visual-gestural modality as it bears on the investigation of brain organization for language. We have now studied intensively more than 20 deaf signers with left or right hemisphere focal lesions; all are highly skilled ASL signers and all used sign as a primary form of communication throughout their lives. Our subjects are examined with a basic neurological exam, neuroradiological studies, and a battery of experimental probes, including formal testing of ASL at all structural levels as well as with spatial cognitive probes [16].

Our results to date have been clear: left hemisphere damaged (LHD) signers revealed marked sign language aphasias that varied depending on lesion site, but none of the right hemisphere damaged (RHD) signers were impaired in any aspect of ASL grammar; their signing was rich, complex and without deficit, even in the spatial organization underlying sentences of ASL, and despite sometimes severe spatial cognitive deficits [2–5, 8, 13, 16]. Many of the sign language deficits we have seen in the LHD signers were not dissimilar to what one would expect had they been hearing–speaking individuals with similar lesions. For example, one subject had a large anterior lesion with a resulting agrammatic aphasia, another subject had a large perisylvian subcortical lesion and was paragrammatic for sign [2, 5, 16], and yet another subject with a temporo-parietal lesion presented with a conduction-like sign aphasia [13]. Some of our cases, however, are hinting at possible differences in the left hemisphere organization for signed language as compared with spoken language (see discussion in Poizner et al. [16]). In this report, we describe a new patient of particular interest in this respect: following a left occipital lesion the patient presented with a highly unusual language profile consisting of a severe sign language comprehension deficit in the face of spared production.

Case LHD-111

LHD-111 was a right-handed deaf woman who suffered a stroke in February, 1981 at the age of 62 years. Her parents were hearing, and she became completely deaf at the age of 18 months. She attended a residential school for the deaf. ASL was her preferred means of communication throughout her life. She married a deaf man and lived within a deaf cultural group where sign language was the primary mode of communication. Before her stroke she had no difficulty reading and writing English, and used a teletype device regularly.

Neurological examination was performed in September, 1983. Her general neurological examination was remarkable for a dense right homonymous hemianopia, a moderate spastic right hemiparesis affecting face, arm and leg, and moderate impairment of pin sensation on the right lower extremity. She used a wheelchair and could aid in transfers. Eye movements, coordination, light touch and position sense, and left-sided strength were intact. CT brain scan without contrast showed what appeared to be a single old ischemic infarction involving the territory of the left posterior cerebral artery. The lucency involved
the medial left temporal and occipital lobes, the left occipital pole, and the white matter that
gave rise to the splenium of the corpus callosum. There was no evidence of abnormal
lucency in the splenium itself or in the midbrain, internal capsule, or thalamus. Throughout
its extent, the left lateral ventricle was enlarged compared to the right (see Fig. 1).

Two years post-stroke she was administered the Salk Sign Diagnostic Aphasia
Examination (SDAE)—our ASL adapted version of the Boston Diagnostic Aphasia
Examination [12]—and a number of experimental probes described below. Her Salk-
SDAE profile of sign characteristics is given in Fig. 2 and contrasted with normal deaf
control profiles and the composite profiles of other left and right hemisphere damaged
signers. As can be seen in the profile, LHD-111’s signing had normal melodic line (rhythm
in the sign stream), phrase length and grammatical complexity. She had no difficulty with
articulatory agility nor sign finding. There were occasional paraphasias, but these were
infrequent. Despite this relatively normal sign performance on production related scales,
her score on the sign comprehension scale was at floor—none of our other subjects have
shown such an extreme comprehension deficit. In what follows we describe her
performance in more detail.

Sign language production. As noted, the patient’s sign language production was fluent,
grammatical and coherent, with a normal balance of content and function signs (or
morphemes). Her signing was thus neither agrammatic nor paragrammatic (see Fig. 2).
The examiner had no difficulty understanding her signing. In contrast, LHD-111
frequently did not understand the examiner’s questions. For example, when asked how
she communicated with her parents, she responded “Before my name was Jones” (English
gloss; a pseudonym was substituted for the actual name). As a further example of LHD-
111’s preserved sign production, consider the following English gloss of her description of
the Cookie Theft Picture (note the failure to comprehend the examiner’s question).

LHD: The boy reached out for the nuts. (To the examiner:) I don’t know what those
are, are they nuts?*
Examiner: Cookies.
LHD: The boy grabbed the cookies and he is falling down. The boy is falling down. He
tried to grab the cookies but missed. He went to grab them but came up empty-handed.
Examiner: What happened to the others?
LHD: The boy took the cookies and fell down (misunderstanding the question).
Examiner: What happened to the mother?
LHD: The mother is day-dreaming washing dishes looking outside. Suddenly, she was
surprised to see that something was wrong. The (paraphasic error for ‘water’) was
overflowing.
Examiner: What happened with the girl?
LHD: The girl screamed and yelled. She yelled.

As this sample demonstrates, her ASL production was essentially intact. Although she did
occasionally produce paraphasic errors, these were rare; to the extent that there was
aphasic symptomatology in her production, it was minimal relative to her comprehension
deficit described in more detail below.

*As we point out below the patient is alexic. It is therefore not surprising that she mistook the cookies for nuts,
because in the Cookie Theft picture it is unclear what is in the container if one cannot read the word ‘cookie’
printed on the side.
Sign language naming. Her naming ability was also preserved. She correctly named six out of six pictures of objects, six out of six pictures of actions, and five out of six colours. She had some difficulty naming letters, typically taking longer to respond on these items, but still she correctly named four out of six.

Sign language comprehension. In a series of tests, we found the patient’s comprehension of ASL to be severely impaired. Her sentence-level comprehension of ASL was virtually absent. She could not follow simple commands such as the ASL equivalent of ‘stick out your tongue,’ or ‘point to the door.’ In further probes of comprehension she made only two correct responses out of 22 on our ASL adaptation of the Token Test [10]. For example, she was able to follow the command POINT TO ANY CIRCLE, but she was unable to follow commands such as POINT TO ANY SQUARE (she pointed to a circle), POINT TO ANY BLACK CIRCLE (she pointed to a white circle) and POINT TO SMALL WHITE CIRCLE (she pointed to a large white square). Her performance on a picture-matching sentence comprehension test, using simple active sentences such as the ASL equivalent of ‘the dog bit the cat,’ was at chance. In contrast her comprehension of single signs, while still far from normal, was less impaired. On the sign discrimination subtest of the Salk-SDAE in which the patient is asked to match signed words to pictures (out of arrays of six), she correctly identified six out of six object stimuli (BED, DOLL, AIRPLANE, TREE, TELEPHONE, BIRD) and five out of six ‘actions’ stimuli (she correctly identified SMOKING, COMBING, SLEEPING, FALLING, CRYING; she failed to identify DRINKING). In another single-sign-to-picture matching task she scored less well, matching only 20 of 30 signs to correct pictures (out of arrays of four), although still significantly better than chance (67% correct > 25% expected by chance; $t(29) = 4.76$, $P < 0.001$). However, one of the three foil pictures was semantically related to the correct picture (e.g. ‘window’ instead of ‘door’ or ‘camera’ instead of ‘binoculars’) and nine of her 10 errors involved the choice of the semantically related foil. If we restrict our statistical analysis to the two semantically related choices (as LHD-111’s responses apparently were)—that is, take chance to be 50% correct—we find that the patient is distinguishing between the correct picture and a semantically related incorrect picture only slightly more than you would expect by chance (67% vs 50% correct; $t(29) = 1.90$, $P = 0.07$). It would seem then, that her access to the meaning of single signs is semantically underspecified, allowing relatively good performance as long as response choices are not too semantically similar.

Sign language repetition. Her performance on a test of repetition of sign varied depending on whether she was asked to repeat single signs or signed sentences. In a test of single sign repetition, she correctly repeated eight of the 10 items presented to her, these were BED, AIRPLANE, WHAT, PURPLE, GREEN, W, 15 and BEAUTIFUL. The two items she failed on were 1776 (on successive attempts with the stimulus repeated for each she responded 1775, 19 and 1773) and CALIFORNIA-GOVERNMENT (she responded CALIFORNIA), both of which are comprised of sign sequences: as it was presented by the examiner, 1776 is composed of three signs, 17, 7 and 6 (it can be reduced to two signs 17 and 76), and CALIFORNIA-GOVERNMENT is composed of the signs CALIFORNIA and GOVERNMENT.

In a test of signed sentence repetition she was severely impaired. She responded correctly on only one out of 16 items. She was, however, able to repeat many of the words in the sentences suggesting that she did have access to the single signs, however it was clear from her response sentences that she was not able to derive the combinatorial meaning of the
Fig. 1. CT scan demonstrating infarction in the left medial occipital and temporal lobes.
Fig. 2. Salk Sign Diagnostic Aphasia Examination profiles of sign characteristics for normal deaf controls, RHD deaf signers (n = 7), LHD deaf signers (n = 9) and the present case LHD-111. Note specifically (i) the scale for sign comprehension (shaded) where LHD-111 is at floor and is far worse than any other LHD signer studied thus far, and (ii) the production related scales (e.g. grammatical form, phrase length, sign finding) where LHD-111 performs just like normals. Note also the relatively few paraphasias and normal sign finding scale, ruling out a Wernicke-like characterization of the patient's aphasia.

signed sentences. For example, when presented with the sign-sentence 'LAST NIGHT ME SEE HIM EAT NUTS' she responded 'YESTERDAY ME GO EAT ICE-CREAM'; likewise when presented with 'ME COME HOME WANT EAT' she responded 'COME, COME EAT WITH ME'. So she appears to understand some of the individual signs and is able to repeat them, but the proposition encoded by the sentence is lost.

Comprehension of manually spelled words. ASL has a manual (fingerspelled) alphabet (one sign for each English letter) that can be used to render proper names, technical terms
and other English words that do not have a corresponding sign. LHD-111's comprehension of manually spelled words, like her comprehension of written words was severely disrupted. She could not identify body parts when prompted with a manually spelled stimulus. She attempted four items, E-A-R, N-O-S-E, S-H-O-U-L-D-E-R and K-N-E-E, failing them all. For example, when asked 'WHERE YOUR E-A-R?' she responded 'B-E-A-R'. In another test, when she was asked to identify manually spelled words by responding with the corresponding sign she correctly identified only three out of nine items. For example, in response to N-O she signed ME and for F-I-F-T-E-E-N she signed EARLY.

English reading comprehension and writing. The patient had significant impairment in reading written English. She was unable to read single words printed on cards. To both the words 'tree' and 'circle' she responded with the sign 'COW', to the word 'telephone' she responded 'HORSE'. She did not attempt any additional items on this test. She was unable to match pictures with printed words (from an array of six pictures for each word), scoring only two out of 10 correct (17% correct is chance-level performance), although she had been able to give the sign for most of the pictured items as noted above. Her sentence-level reading was also impaired in both a reading 'aloud' task (where she was asked to provide the signs for the words as she was reading) and in a multiple choice sentence completion task. A previous neurological exam indicated that she was able to write single words but had difficulty with sentences. During our testing she refused to write anything besides her name.

In summary, LHD-111 presented with severe impairment of sign comprehension, with near normal sign production. She also had significant impairments of English reading comprehension, and comprehension of manually spelled words. We can rule out the possibility that LHD-111's comprehension problems are secondary to visual acuity limitations on the following grounds. First, as noted above, her comprehension of single signs was relatively preserved; if visual acuity were at issue, one would expect equal difficulties on single signs. Second, she was able to recognize pictures well enough to name them in sign and had no difficulty parsing a relatively complex scene such as the Cookie Theft picture. Finally, she performed well on the visual-spatial tasks she was given; she was able to copy a cross and a triangle (although she had difficulty with a cube), and she performed in the normal range on the Benton Facial Recognition Test [6].

DISCUSSION

The most striking aspect of LHD-111's syndrome was the almost total loss of ASL sentence-level comprehension in the face of grammatically complex and appropriate production. She was unable to comprehend even simple ASL sentences such as the equivalent of 'the dog chased the cat' and could not follow simple one-step ASL commands. In marked contrast to her severely impaired comprehension, LHD-111's ASL production was fluent, grammatically complex and coherent, she was neither agrammatic nor paragrammatic. Her responses to pictured stimuli were appropriate to the task and displayed a normal range of ASL syntactic structures. As part of her description of the 'Cookie Theft' picture, for example, she produced the ASL equivalent of 'He went to grab the cookies but came up empty-handed'. Yet when she was presented with sentence-level sign stimuli, she performed no better than chance, even with the simplest ASL sentence stimuli or ASL commands. Even her single sign comprehension was impaired to some
degree. Importantly, as argued above, her comprehension deficit cannot be attributed to visual acuity or general perceptual limitations, rather it appears to be linguistic specific.

The wide discrepancy between comprehension and production capacity strongly suggests that this is not a Wernicke's aphasia-like syndrome because one expects a significantly greater production deficit, involving more than just occasional paraphasias and also paragrammatic output. Rather, her symptomatology more closely resembles a disconnection syndrome. Specifically, we propose the following model. Perisylvian regions of the left hemisphere are crucially involved in processing sign language as well as spoken language; this has been demonstrated in a number of cases and is a crucial piece of information in accounting for the present deficit [2]. In addition, we propose that the pathway for sign stimuli perceived in the left visual field crosses from the right visual cortex via projections across the splenium of the corpus callosum to corresponding visual areas of the left hemisphere; these areas then project to the language regions of the left hemisphere. In LHD-111's case, her lesion involved the left visual cortex and white matter adjacent to the splenium of the corpus callosum and thus disconnected the intact right visual cortex from the intact language areas in the left hemisphere. A severe sign language comprehension deficit with spared production was the result. This is strong evidence for a left hemisphere organization of sign language because the right hemisphere is clearly unable to support ASL comprehension.

This model is analogous to the model proposed for alexia without agraphia, a well documented disorder in which non-aphasic patients are severely impaired in comprehending written language, but unimpaired in the ability to write [11] (and references therein), and indeed LHD-111 also presented with alexia. There does appear to be a difference in the degree to which sign comprehension and written-word comprehension are affected, however. While the patient was unable to read single words, she was able to understand many isolated signs. Her single sign comprehension was certainly not without error, but even where she did respond incorrectly to a sign stimulus, her errors were often semantically related to the target. The etiology of this difference between reading words and perceiving signs is unclear. Her word-reading deficit is characteristic of pure alexia, and it is reasonable to expect a parallel deficit in single-sign comprehension, yet clearly the latter is relatively preserved. At the same time, her strong tendency to make semantically related errors in one task of sign comprehension is reminiscent of the kinds of errors seen in deep dyslexia, a syndrome which has been attributed by some to right hemisphere language ability [7, 9, 17, 18]. Perhaps LHD-111's single sign comprehension represents the right hemisphere's capacity for processing ASL. Why the right hemisphere is less efficient at reading words than signs remains a puzzle (see Baynes [1] for discussion of related issues).

One additional finding requires discussion, namely, the fact that LHD-111 did produce occasional paraphasic errors. Such errors would not be expected following a left posterior cerebral artery (PCA) infarct in a hearing patient. Perhaps her paraphasic errors can be attributed to limited left thalamic involvement that was not apparent on her CT scan. This seems unlikely however given the rarity of thalamic involvement in PCA infarcts, and the lack of aphasic symptomatology in those cases with such involvement [15]. An alternative explanation is that in deaf signers some degree of neural reorganization takes place as a result of the visual input of linguistic information. Thus, one might expect an increased
involvement of posterior regions of the left hemisphere in some language-related neural systems. More cases are needed in order to map out the neuroanatomy and plasticity of these systems.

In summary, we have described the case of a deaf signer with a severe sign comprehension deficit and spared production following a lesion in the distribution of the left PCA—a lesion that would not be expected to cause aphasia in a hearing patient. We suggest that this is a case of 'sign blindness' due to a disconnection between visual information in the intact right visual cortex and the intact language areas in the left hemisphere. This case provides some of the strongest support to date for the modality independence of left hemisphere dominance for language. However, this case also suggests important differences in the neural organization of signed vs spoken language within the left hemisphere.

Acknowledgements—This research was supported in part by National Institutes of Health grants R01 DC00201; R01 DC00146; R37 HD13249; P01 NS19632; as well as National Science Foundation grant BNS 8911486. We are grateful to Hanna and Tony Damasio, Jennings Falcon, Maureen O'Grady-Hines, Howard Poizner, Kathleen Say, Dennis Schemenauer and Mark Williams for their help in these studies, and to Eleanor Safran for helpful comments on an earlier draft. We are also grateful to the subjects and their families for participating in these studies. Illustration, copyright Ursula Bellugi, The Salk Institute for Biological Studies, La Jolla, CA, U.S.A.

REFERENCES