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David Corina & Nicole Spotswood, Davis, California (USA)

32. Atypical signing

Abstract

Similarities and differences between impairments across modalities can help answer the question of whether sign language and spoken language are processed identically and independently of the perceptual and articulatory channels in which they are instantiated, or whether they reflect how language is shaped by modality. This chapter begins with a

brief definition and overview of the notion of 'atypical sign language'. The discussion of acquired impairments briefly reviews the neurolinguistics of language impairment, and the contribution of various abilities/impairments outside the 'language module'. It then considers motor (including motor planning), linguistic, cognitive, and neuro-psychiatric impairments. Within the field of motor impairments, disorders such as Parkinson's disease and other degenerative conditions are considered in terms of their impact on the articulation of signing. Linguistic impairments arising from stroke are dealt with in case studies and group reports of left- and right-lesioned signers, in relation to modality-dependence/independence issues. Within the field of developmentally atypical signing, various individual and small group studies are presented and discussed in depth: Specific Language Impairment, Williams syndrome, Down syndrome, Autistic Spectrum Disorder, Tourette's syndrome, and Landau-Kleffner syndrome. The chapter also includes a brief discussion of the impact on sign language of neuropsychiatric impairments such as dementia and schizophrenia and concludes with consideration of whether language impairments reside in a specific modality or are modality-independent.

1. Introduction

Since the early studies of Broca in the 19th century, insights into the brain and its functions have come from the study of individuals with language and communication impairments. When language is not acquired effectively, or there is a breakdown in skills, then identifying the sources of impairment offers crucial insights into structure and processing. In the context of sign language, studies of atypical language provide a unique window on the relationship of language and modality (Woll/Morgan 2011). Similarities and differences between impairments across modality can help answer the question of whether sign language and spoken language are processed identically and independently of the perceptual and articulatory channels in which they are instantiated, whether they reflect how language is shaped by modality, and the extent to which language relies on other cognitive processes.

Interest in atypical signing goes back much further than contemporary readers might suppose. In the first issue of *Brain* (1878), John Hughlings Jackson commented "No doubt by disease of some part of his brain the deaf-mute might lose his natural system of signs" (p. 328), and there are several accounts of impairments in signers following stroke throughout the remainder of the 19th and the first half of the 20th centuries. Grasset (1896) described a patient who had developed an impairment in sign language following a stroke which was not attributable to motor impairment of his right arm, concluding "he really is, therefore, genuinely aphasic in the right hand, in the true and only meaning of the word". Other early authors include Burr (1905) and Leischner (1943). Leischner analysed a congenitally deaf trilingual (sign language, Czech, and German) patient following a stroke affecting the lower left parietal and superior temporal lobes. He described impairments in his patient's oral, written, and sign language.

Critchley (1938) describes acquired impairments in fingerspelling in the case of W. H. H., aged 42 years, who had learned fingerspelling and lip-reading after becoming deaf in childhood, but who also had some speech. Within four weeks of his left hemi-

sphere stroke, power in the right arm and speech were restored. He could not write, calculate, or fingerspell (although his ability to understand these symbols did return). Interestingly, the initial examination of the patient was assisted by the chaplain to the school for the deaf. Despite partial recovery, confusion between -A- and -E- is reported (in the British two-handed manual alphabet, these are articulated by touching with the dominant index finger the index and middle fingers of the non-dominant hand respectively). His fingerspelling was also described as telegraphic and lacking grammar; he could not recite a list of vowels or the alphabet, although he could write, type, and read.

There was also early interest in apraxia (the inability to execute a voluntary motor movement despite being able to demonstrate normal muscle function) of signing and the dissociation between apraxia and aphasia in signers as in speakers. Critchley (1938) and Tureen, Smolik, and Tritt (1951) both state that there was no apraxia in their signing aphasics. Leischner's patient (1943) was aphasic but not apraxic: he could perform a series of movements to command requiring the manipulation of real objects, for instance, light a candle with a match, take glasses off and put them into their case, etc. Sarno, Swisher, and Sarno (1969) report that their patient could imitate correctly the movements of using a toothbrush, using a pencil, sweeping, etc., and also that he could correctly manipulate objects such as a comb and a razor, and thus describe him as not showing "non-language apraxia". Kimura, Battison, and Lubert (1976), in the first modern study of a signer following a stroke, explored both aphasia and apraxia. Although not impaired on the usual apraxia tests, he was impaired, relative to non-aphasic deaf controls, in the imitation of complex non-linguistic hand movements, suggesting the need for specialised assessments of apraxia in signers.

Leischner (1943) was unwilling to label impairments in signing as aphasia and proposed the term "asymbolia" instead, although Douglass and Richardson (1959) supported the concept of a "language zone" in the dominant hemisphere of the congenitally deaf, comparable to that for spoken language in hearing people. Sarno, Swisher, and Sarno (1969) were the first to conclude that "aphasia in the congenitally deaf is entirely equivalent to that in normal hearing people". Although this perspective has largely been accepted, there has been recent interest in whether there are indeed modality-related differences (see section 5).

Apart from aphasia and apraxia, there has been relatively little interest until recently in developmental and acquired impairments of sign language. Following a detailed discussion of impairments following stroke below, a range of impairments in sign language will be discussed. For the most part, these are case studies or small group studies.

2. Acquired impairments in sign language

2.1. Impairments resulting from stroke

2.1.1. Left hemisphere damage

Case studies of deaf signing individuals with acquired brain damage, and neuroimaging studies of healthy deaf subjects, have provided confirming evidence for left hemisphere

dominance in processing sign languages as well as spoken languages (see chapter 31, Neurolinguistics). Deaf signers, like hearing speakers, exhibit language disturbances when left-hemisphere cortical regions are damaged (e.g., Hickok/Love-Geffen/Klima 2002; Marshall et al. 2004; Poizner/Klima/Bellugi 1987; for a review, see Corina 1998a,b and MacSweeney et al. 2008). In addition, there is good evidence that cerebral organisation in deaf signers within the left hemisphere follows the familiar anterior/posterior dichotomy for language production and comprehension that is found in spoken language users (see Figure 31.1 in chapter 31). Right hemisphere damage, although it can disrupt visual-spatial abilities (including some involved in sign language processing), nevertheless does not produce sign aphasia (Atkinson et al. 2005).

Poizner, Klima, and Bellugi (1987) present the first modern set of studies of signers (of American Sign Language, ASL) with aphasia arising from left hemisphere lesions. In relation to production, damage to the left anterior frontal lobe (Broca's area) is associated with aphasia in signers as it is in speakers. For example, Poizner, Klima, and Bellugi (1987) report on patient G. D., who had a large lesion in Broca's area and who had aphasia, with dysfluent single-sign agrammatic utterances, but unimpaired comprehension. She also produced reduced linguistic facial expression relative to affective expression (Corina/Bellugi/Reilly 1999).

In relation to comprehension, the left temporal cortex plays an important role in sign language as it does in comprehension of spoken language. Hickok, Love-Geffen, and Klima (2002) and Atkinson et al. (2005) compare the sign language comprehension abilities of left- and right-hemisphere damaged ASL and British Sign Language (BSL) signers respectively. Signers with left-hemisphere posterior temporal lobe damage were found to perform worst, exhibiting significant impairments on single sign and sentence comprehension. Fluent aphasia − impaired language comprehension accompanied by fluent production, although often with semantic and phonological paraphasias − has been reported in both spoken language and sign language. Although less common than aphasias with dysfluencies, cases are reported by Chiarello, Knight, and Mandel (1982), Poizner, Klima, and Bellugi (1987), and Corina et al. (1992).

2.1.2. Right hemisphere damage

Typically, signers with damage to the right hemisphere are reported as having well-preserved language skills, even when they exhibit visual-spatial deficits (Poizner/Klima/Bellugi 1987; Atkinson et al. 2004). This resembles findings for hearing non-signers but is surprising in view of the role of vision and space in sign language processing. The case study of J. H., a deaf signer with a right hemisphere lesion involving frontal, temporal, and parietal regions (Corina/Kritchevsky/Bellugi 1996), presents a striking example of profoundly impaired visual-spatial skills but preserved sign language abilities.

Despite the absence of aphasia in right hemisphere-lesioned signers, there is evidence that right hemisphere structures may play a crucial role in the production and comprehension of classifiers and in the processing of non-manual linguistic elements. In tests requiring classifier descriptions, Poizner et al.'s (1987) subject D. N. showed problems in the depiction of movement direction, object relations, and object orientation. In situations where two-hand articulations were required, D. N. displayed great hesitancy and often made multiple attempts to correctly represent the spatial relation-

ship of the referents, although D. N. had no motor weakness to account for these errors. Although syntactic processing is generally intact in right hemisphere-lesioned signers, in contrast to the common disturbances of syntactic processing in left hemisphere-lesioned signers, there are reports of signers with right hemisphere damage exhibiting syntactic problems. Subjects S. M. and G. G. (right hemisphere-damaged subjects tested by Poizner/Klima/Bellugi 1987) performed well below controls on two tests of spatial syntax. Atkinson et al. (2005) report on the difficulties experienced by right hemisphere-lesioned signers on tests of comprehension of spatial syntax involving locative sentences and classifiers.

As is the case with hearing individuals (Marini et al. 2005), right hemisphere damage in signers may result in disruption of discourse abilities. One problem identified for right hemisphere-lesioned signers is in the processing of non-manual linguistic elements (Corina/Bellugi/Reilly 1999; Atkinson et al. 2004). Atkinson and colleagues report a group study comparing processing of manual and non-manual negation in BSL in right- and left-lesioned signers (see chapter 15, Negation, for details). Both groups performed comparably to controls in processing manually marked negation, but right hemisphere-lesioned signers performed below chance on negation marked non-manually through head shake and facial expression only. The authors analyse non-manual negation as prosodic at surface level and attribute the failure of right hemisphere-lesioned signers to the difficulties faced by this group with prosodic processing generally.

Other types of disturbance in discourse are reported in two right hemisphere-lesioned patients: J. H. (Corina/Kritchevsky/Bellugi 1996) and D. N. (Emmorey/Corina 1993; Emmorey/Corina/Bellugi 1995; Poizner/Kegl 1992). These two cases reveal contrasting impairments. J. H. showed occasional non sequiturs and abnormal attention to detail in signing and picture description tasks, behaviours that are typically found in the discourse of hearing patients with right hemisphere lesions. D. N. showed a different pattern of discourse disruption; her within-sentence use of spatial indexing was unimpaired, but her use of space across sentences was inconsistent. In order to maintain intelligibility, D. N. used a compensatory strategy in which she restated the noun phrase in each sentence, resulting in a repetitive discourse style. The cases of J. H. and D. N. suggest that right hemisphere lesions in signers can differentially disrupt discourse content (as in the case of J. H.) and discourse cohesion (as in the case of D. N.).

2.1.3. Differences between spoken language and sign language

Although the above studies suggest that comparable impairments in spoken language and sign language arise from lesions in the same locations, there is some controversy in regard to the degree of anatomical overlap observed in comprehension problems in spoken and sign languages. These in turn have been related to existing questions about the mechanisms involved in some aspects of sign language, in particular the relationship between gesture and linguistic structure. The fact that linguistic forms in sign languages exploit visual-gestural systems rather than the auditory-oral systems of spoken language leaves open the possibility that there may be different neural systems underlying language in these two modalities.

It is noteworthy that the cases described by Chiarello, Knight, and Mandel (1982) and Poizner, Klima, and Bellugi (1987) and the case study described by Corina, Vaid, and Bellugi (1992), exhibited fluent aphasia with severe comprehension deficits. Lesions in these case studies did not occur in Wernicke's area, but rather involved more frontal and inferior parietal areas. In these cases, lesions extended posteriorly to the supramarginal gyrus. This is notable, as lesions associated with the supramarginal gyrus alone in users of spoken language do not typically result in severe speech comprehension deficits. These observations have led some to suggest that sign language comprehension may be more dependent than speech on left-hemisphere inferior parietal areas (regions associated with somatosensory and visual motor integration) (Leischner 1943; Chiarello/Knight/Mandel 1982; Poizner/Klima/Bellugi 1987; Corina 1998b) while spoken language comprehension might rely more heavily on posterior temporal association regions whose input includes networks intimately involved with auditory speech processing (see section 4.2 below on Landau-Kleffner syndrome). As Corina and Spotswood discuss in chapter 31, neuroimaging research also points to modality-conditioned effects on neural processing of sign languages (Newman et al. 2002; Emmorey et al. 2002; Emmorey et al. 2005; MacSweeney et al. 2006).

A second issue is whether the functional mechanisms required for comprehension of sign language and spoken language are identical. Many sign languages express locative relationships and events with spatialised structures and have inventories of highly productive grammatical forms, classically referred to as classifiers or classifier predicates (see chapter 8 for discussion), which participate in these constructions. Liddell (2003) has argued that such constructions are partially gestural in nature. The theoretical status of such spatialised grammar is a subject of debate which has important implications for the understanding of the neurolinguistics of sign language. Dissociations between gesture and sign language abilities have been reported in a case study of an aphasic signer (Marshall et al. 2004) suggesting that these are separate. However, several studies have found differential disruptions in the use and comprehension of sentences that involve usage of spatialised grammar compared to other grammatical constructions. For example, Atkinson and colleagues (2005) conducted a group study of left and right hemisphere-damaged signers of BSL. They devised tests that included single sign and single predicate-verb constructions (e.g., THROW-DART), simple and complex sentences that varied in argument structure and semantic reversibility, locative constructions encoding spatial relationships and constructions involving lexical prepositions, and a final test of classifier placement, orientation, and rotation. Their findings indicated that left hemisphere damaged (LHD) BSL signers, relative to elderly control subjects, exhibited deficits on all comprehension tests. Right hemisphere damaged signers (RHD) did not differ from controls on single sign and single predicate-verb construction, or on sentences that ranged in argument structure and semantic reversibility. RHD signers (like LHD signers), however, were impaired on tests of locative relationship expressed via classifier constructions and on a test of classifier placement, orientation, and rotation.

One interpretation is that the comprehension of these classifier constructions requires not only intact left hemisphere resources, but intact right hemisphere visual-spatial processing mechanisms as well. That is, while both LHD and RHD signers show comprehension deficits, the RHD signers' difficulties stem from more general visual spatial deficits rather than linguistic malfunction per se. The question of whether these

visual spatial deficits are "extra-linguistic" lies at the heart of the debate. Atkinson et al. (2005) suggest that the deficits stem from the disruption of processes which map non-arbitrary sign locations onto real-world spatial positions. However, as Corina (personal communication) has pointed out, such forms are not only used to refer to real-world events, but imaginary and non-present events as well. The broader point he makes is whether *aphasic* deficits should be solely defined as those that have clear homologies to the left hemisphere impairments that are evidenced in spoken languages, or whether the existence of sign languages will force us to reconsider the concept of linguistic deficits.

2.1.4. Paraphasia

In spoken language, the substitution of an unexpected word for an intended target is known as paraphasia (Corina 2000). Most paraphasias have a clear semantic relationship to the target. Several reports of semantic paraphasia in sign languages can be found in the sign aphasia literature (Poizner/Klima/Bellugi 1987; Brentari/Poizner/Kegl 1995; Corina/Vaid/Bellugi 1992; Marshall et al. 2004). Poizner et al.'s (1987) subject P.D., for instance, substituted BED for CHAIR, and DAUGHTER for SON; "Charles" (Marshall et al. 2004) substituted CAR BOX for LORRY.

Paraphasias can also involve phonological substitution, sometimes resulting in a non-word or a word related in form but not meaning. Phonological paraphasias are also found in signers. Although they affect all the major formational parameters of sign languages (see chapter 3, Phonology), the distribution of paraphasic errors among the four parameters of sign formation appears to be unequal: handshape configuration errors are the most widely reported, while paraphasias affecting movement, location, and orientation are less frequent. An unusual case of sign paraphasia, resulting from damage to the left frontal operculum, is reported by Hickok et al. (1996). R. S. experienced an initial expressive aphasia that largely resolved, however with lingering problems of word finding and frequent phonemic paraphasia. Corina has pointed out the noteworthy nature of these paraphasias, which demonstrate how language modality may uniquely influence the form of linguistic deficit – in this case, impairments with no clear parallels to spoken language disruption (see section 5 below for further discussion of the implications of these and other studies for understanding the relationship between language and modality).

Poizner, Klima, and Bellugi (1987) speculated that the perceptual processing involved in the comprehension of spatialised syntax involves both left and right hemispheres; certain critical areas of both hemispheres must be relatively intact for accurate performance. The syntactic comprehension deficits found in right and left hemisphere-damaged subjects raise an interesting theoretical question: are these deficits aphasic in nature, or are they secondary impairments arising from a general cognitive deficit in spatial processing? Further work is required to tease apart these complicated theoretical questions.

2.2. Parkinson's disease

Several studies in the modern era have addressed the issue of motor linguistic dissociations from general motor functions. One such strand of research concerns the linguistic

behaviour of signers with Parkinson's disease (PD). The main symptoms of PD result from the death of dopamine-generating cells in the substantia nigra, a region of the midbrain. PD patients reveal qualitatively different impairments from those found in aphasic signers. The motoric aspects of sign language production are greatly disrupted by PD, although language is preserved (Brentari/Poizner 1994; Brentari/Poizner/Kegl 1995; Poizner/Kegl 1992, 1993). For example, PD signers typically show a dampening of facial expression and reduction of the amplitude of movement in general (Loew/Kegl/Poizner 1995). In addition, the errors produced by signers with PD are more pronounced at sentence and discourse level than in signs produced in isolation (Poizner 1990). Thus the deficits exhibited by the signers with PD reflect, in large part, an impairment in the production of complex movement sequences (Tyrone/Woll 2008). Brentari, Poizner, and Kegl (1995) demonstrated that signers with PD showed marked disturbances in the temporal organization and coordination of the two motor subsystems of the ASL sign stream: handshape and movement. The change in handshape that is required for transition from one sign to another normally occurs early in the transition between signs in control signers; that is, the normal timing relationship between handshape formation and arm movement is quite asynchronous. Signers suffering from PD, however, largely synchronized the change in handshape with the start and end of the movement. They thus reduced the temporal variability in the relationship between the handshape and movement motor subsystems of ASL. These sorts of changes parallel those found in non-linguistic manual activities of hearing individuals with PD.

2.3. Progressive supranuclear palsy

Progressive supranuclear palsy (PSP) is a progressive neurological condition with some surface similarities to PD, although affecting different areas of the brain, mainly the rostral brainstem and its projections to the basal ganglia, cerebellum, and cerebral cortex. Articulation difficulties in hearing patients typically emerge early in the course of the disease and disrupt several aspects of speech. The characteristic features of PSP are articulatory incoordination and palilalia − the repetition of entire words at decreasing amplitudes without pause. The only case study of a signer with PSP (Tyrone/Woll 2008) reported frequent deviations from citation form, with errors in orientation, handshape, and location, poor coordination, atypical repetitions, and involuntary movements. Most striking was the finding of palilalia. The disrupted signing can be considered as palilalic because entire signs were repeated, and repetitions became smaller in movement amplitude, parallel to palilalia in speech. Research on the forms of articulatory deficits across language modalities can lend insight into the basic articulatory units of both sign and speech. This study suggests that articulation is a modality-independent function; it consists of the rapid, complex, sequential, coordinated movements necessary for language production. Moreover, preliminary findings suggest that the same neural structures govern articulation in both speech and sign (but see section 4.3).

2.4. Dementia

Despite the prevalence of dementia in the general population, there have been virtually no studies of dementia in the Deaf community. A small recent study (DiBlasi

2011) explored changes in Deaf individuals' ASL as their cognitive status declines. This study involved five individuals with cognitive impairments and five controls. They were assessed on the Mini-Mental State Examination adapted for use with signers (Dean et al. 2009). Each participant also was given two minutes to describe the Cookie Theft picture (see Poizner/Klima/Bellugi (1987) for an illustration). Their discourse was analyzed for number of utterances, number of words per utterance, use of phonology, morphology, and syntax, and content. There were significant differences between patients and controls on a number of features, including number of signs per utterance, and occurrence of phonological errors. A larger-scale study is currently under way with BSL signers in the UK, which is collecting data on normal cognitive and linguistic function in signers aged between 50 and 90, in order to develop a cognitive and linguistic screening tool for use with the signing population (Atkinson et al. 2011).

3. Developmental impairments in sign language

The study of children exposed to a sign language but following an atypical course of development because of developmental disorders is a very new area of study. In this population, the issues are particularly complex and involve questions of whether language development is affected in modality-specific or modality-independent ways. In cases where non-verbal cognitive deficits are present, the impact of these on the acquisition and use of a language perceived and produced in the visual-spatial modality must be considered. Typically, deafness is an exclusionary criterion for studies of language impairment because these studies have always focused on the acquisition of a spoken language. The inclusion of children exposed to sign languages but presenting with atypical development has the potential to open a new window on the question of whether language impairments originate from deficits in the cognitive, linguistic, or perceptual systems.

Several types of studies are reviewed here. Some concern hearing children and young people who are atypical speakers of English and who also use BSL, for example, hearing identical twins with Down Syndrome, who are the children of deaf parents. Other cases discussed in section 4 include a hearing young man with Landau-Kleffner Syndrome, aphasic in English but with relatively good BSL; and Christopher, a linguistic savant who learned BSL as an adult despite cognitive and language impairments. The remainder of this section concerns, deaf children and young people. Cases include a young deaf woman with developmental visual-spatial impairments (Williams Syndrome) and children with Specific Language Impairment, autism, and Tourette's syndrome.

3.1. Specific language impairment

Specific language impairment (SLI) is diagnosed where a deficit in normal spoken language acquisition – typically, difficulty with the acquisition of phonology and morphosyntax – is found with no apparent cognitive, social, or neurological cause (Leonard 1998). All theories of SLI attempt to explain the disproportionate difficulty with

phonology and grammar found in SLI but differ in whether they posit a deficit at the level of language or general cognitive processing. Since hearing loss is specifically excluded in diagnosing SLI, deaf children are never included in studies of SLI and there has been relatively little consideration of whether a child exposed to sign language could have SLI (Morgan 2005).

Recently, group studies of SLI have been undertaken in the US (Quinto-Pozos/Singleton 2010; Quinto-Pozos/Forber-Pratt/Singleton 2011) and the UK. Morgan and colleagues (Mason et al. 2010; Morgan/Herman/Woll 2007) have studied SLI in children whose first language is BSL, and who were referred for assessment by teachers or speech and language therapists because of concerns about their sign language development in comparison with their peers. All children had been exposed for at least four years to native signers of BSL and had normal motor and cognitive development. However, on assessments of BSL receptive grammar and grammatical and pragmatic skills (Herman/Holmes/Woll 1999; Herman et al. 2004), 7 of 13 children displayed impaired receptive grammar, and 8 had impaired productive grammar. It is clear that complex morphology is generally impaired in this group study; the results also indicate different profiles of impairment in individual children: affecting phonology, receptive grammar, productive grammar, pragmatics, and discourse. These findings suggest that SLI affects language acquisition in similar ways in both the spoken and signed modalities but that language typology also influences which aspects of linguistic structure are more or less intact.

Morgan and colleagues also report on a related single case study of "Paul", a deaf native signer aged five years, referred for assessment by his school because of worries about his BSL development, which was described as being unusually slow for a native signer. This case is of particular interest, since his impairments cannot be explained by poor input. Paul scored well below the mean for BSL grammar on the BSL Receptive Skills Test (Herman/Holmes/Woll 1999), with success on some difficult items, failure on many easier ones, and with a particularly poor profile on negation, spatial verbs, and classifiers.

The deaf children with SLI show comparable impairments to those found in hearing children with SLI. In both the group study and the study of Paul, impairment was found for grammatical constructions involving verb agreement (see chapter 7). In contrast to typically developing native signers of Paul's age, who use inflectional morphology on the verb GIVE to indicate agent and recipient, as in MAN LETTER GIVE$_3$ ('The man gives the letter to him/her'), Paul signed a sequence of uninflected signs when asked to describe a picture of a man giving a letter to a boy despite prompting, as illustrated in (1) (P = Paul, A = Deaf adult).

(1) P: GIVE GIVE SQUARE GIVE (citation forms) [BSL]
'Give, give the square thing, give.'

A: SQUARE GIVE WHO?
'Who gives the square thing?'

P: GIVE GIVE INDEX(picture) LETTER
'Give, give, (point), letter.'

A: PICTURE WHAT?
'What is in the picture?'

P: LETTER INDEX(picture)
'A letter (point).'

Paul's difficulty in using BSL verb morphology may be linked to the nature of meaning-form mappings using this type of verb in BSL. Signers must simultaneously encode both the core meaning, for instance, using the appropriate handshape for 'giving', and the direction of movement, encoding the identity of the agent and recipient. This packaging of information into a single unit with several components requires good language skills. The sets of data from Paul and from the group study suggest that SLI affects verb morphology in similar ways in sign language (BSL in this case) and spoken language. Where this is the case, sign language data cannot help us to decide whether difficulties with grammatical rules originate from domain-general impairments in information processing which would affect rule learning underpinning language but also other complex systems (Kail 1994) or from a domain-specific linguistic impairment (van der Lely 2005), but the data do suggest that modality-related processing difficulties cannot be the source of SLI.

3.2. Williams syndrome

Early studies of language in Williams syndrome (WS) reported dissociations between profound visual-spatial deficits and impressive receptive and productive language skills (see e.g. Bellugi et al. 1988). While these early studies suggested that language in WS was intact, recent research has been more sensitive to patterns of relative strengths and weaknesses across domains, and it has become clear that language is not wholly intact in WS. A new picture has emerged which suggests that language should be viewed as relatively spared rather than normal (see e.g. Karmiloff-Smith 2008).

English speakers with WS show subtle linguistic impairments that may be related to problems with visual-spatial cognition. Studies of WS in populations speaking languages other than English indicate patterns of impairment in grammar, as the relatively limited extent of morphological marking in English may mask processing difficulties. Volterra et al. (1996) found that Italian speaking subjects with WS produced ungrammatical, or grammatical but atypical, constructions in sentence repetition and story description tasks and made frequent preposition errors. These findings suggest either that language impairments may arise from impairments in visual-spatial cognitive domains or that spatial aspects of both cognition and language are controlled by a higher-level representational system.

The case of a signer with WS is thus of interest since there is the possibility of a more transparent interaction between visual-spatial abilities and language. Atkinson, Woll, and Gathercole (2002) report on the case of "Heather", a young deaf woman. She is of short stature, with a facial appearance and behavioural profile characteristic of WS. Heather uses BSL as her preferred method of communication, although she has some limited ability to lip-read and use spoken and written English. She lives independently in sheltered housing for Deaf people with additional disabilities and regularly attends local Deaf clubs and mixes in the Deaf community. Her command of BSL is strikingly different from her Deaf intellectual peers living in the same sheltered accommodation, in terms of fluency and complexity. However, although not immedi-

ately apparent in spontaneous conversation, she does make consistent errors in her use of some features of BSL.

Heather has clear impairments in non-language visual-spatial ability, measured on a variety of standardised tests. She also displays marked problems with comprehension and production of BSL on standardised assessments, with particular difficulty with spatialised syntax and other grammatical structures using space for grammatical purposes. At sentential level, Heather's production of spatial verbs shows consistent impairment in spatial representations. In her spontaneous signing, she appears to try to deal with her difficulties by choosing English-like structures and a fixed sign order resembling English. For example, Heather uses the prepositions UNDER, ON, and IN rather than classifiers located in spatial relationships to each other to incorporate information about referents and the spatial relationships between them. In general, Heather avoids using classifiers and prefers to use an undifferentiated point with her index finger to locate referents in space. Where she does use classifiers, these are often bizarre (see Atkinson/Woll/Gathercole (2002) for full details).

At the discourse level, Heather also has difficulties with ensuring maintenance of topographic locations across sentences. The results from all the BSL assessments show a disruption in the use of space within BSL, while linguistic devices which do not incorporate spatial relationships, such as noun-verb distinctions and negation, are preserved.

Heather's language abilities in general are well in advance of her visual-spatial abilities. However, her language profile differs from that of hearing individuals with WS, since the subtle impairments found in spoken language in WS are far more transparent in BSL. Most strikingly, for Heather there is a clear dissociation between grammar that relies on space, and grammar that can be specified lexically (e.g. plurals, static locatives). This suggests that although the learning of a visual-spatial language is not in itself dependent on intact visual-spatial cognition, the pattern of breakdown in BSL abilities indicates a dissociation within BSL grammar between devices that depend on grammatical processes involving space and those that do not. Heather's command of grammar appears well preserved except where spatial relationships are conveyed directly. In the latter circumstances, visual-spatial impairment overrides general grammatical ability.

3.3. Down syndrome

Although signs are widely used to support spoken language development in children with Down syndrome (DS), there is only one case study of native sign language development in this population (Woll/Grove 1996; Woll/Grove/Kenchington 1998). Ruthie and Sallie are monozygotic twins. Both parents are deaf and members of the Deaf community. In the presence of their parents and other deaf people, the twins mostly use BSL without voice, although in such contexts, they occasionally address English-only utterances to each other (these appear to function as private asides). In the presence of hearing children and adults and when playing with each other, they use English. As is usually the case in DS, assessments of the twins' verbal and nonverbal ability show that their nonverbal cognitive skills are in advance of their verbal skills. On measures of comprehension of English vocabulary and grammar, at 10 years of age,

they were functioning at a 3 to 4 year old level. Overall, as might be expected, visual and motor skills are relative strengths for both girls.

The twins showed relatively higher skills for BSL vocabulary comprehension than for English. The lexical advantage for signs was not seen in morphology. Sallie's BSL grammar is more advanced than Ruthie's, but neither Sallie nor Ruthie has mental-age appropriate mastery of BSL. Some of Sallie's and many of Ruthie's responses omit spatial relationships completely; in others, they use lexical signs such as IN-FRONT and ON (i.e. English-like structures), rather than representing spatial relationships directly. Across various areas of morphosyntax in both BSL and English, they have difficulty with those with the greatest complexity. For example, they are very good at English plurals which are relatively simple, but poor at those BSL plurals which require classifiers + distributional morphemes, or those with three-dimensional representations of space. Ruthie and Sallie thus find the grammatical system of a sign language no easier to master than that of a spoken language.

Where problems relate to linguistic, as opposed to more general cognitive abilities, delays and difficulties should be seen in language, regardless of modality. The twins' BSL grammar is at a comparable level to their English grammar. This suggests a supra-modal linguistic deficit, with the pattern of varying competences in both languages related to the complexity of the required linguistic devices. This is consistent with observations of difficulties in the acquisition and generalisation of rules affecting complex sentence structure in children with DS. However, some studies of children with DS indicate that although their visual-spatial skills are generally more advanced than their auditory-vocal skills, there may be impairments in the area of spatial representation (Uecker et al. 1993; Vallar/Papagno 1993) and this may suggest differences in the sources of their difficulties in the two languages. In particular, their difficulties in BSL grammar cluster around hierarchically complex structures of a type not found in non-linguistic spatial cognition. Additionally, unlike the relative similarities in grammar cross-modally, their BSL vocabulary is an area of strength compared to English. This in turn raises further questions about the nature of the sign lexicon in terms of such issues as iconicity and phonological structure.

3.4. Autism

There is extensive research on deaf children in cognitive areas such as Theory of Mind and face processing, where norms differ from those in the hearing population, and studies looking at the use of signing with hearing children who have Autistic Spectrum Disorder (ASD) (see Bonvillian (2002) for a review), but relatively few studies looking at deaf children with ASD. The signing of children with ASD is of particular interest because of the ways that some of the known impairments associated with autism are likely to interact with sign language. These include the requirement for signers to understand the visual perspectives of others and the need to look to the interlocutor's face to communicate, in relation to which hearing children with ASD are often reported to be particularly poor. These difficulties are associated with failure to master Theory of Mind, which is delayed in autism. Two recent studies, however, have explored visual perspective and face information processing in deaf children with ASD, respectively (Shield 2010; Denmark 2011).

Shield compared 25 native signing deaf children and adolescents with autism with a control group of 13 typically-developing deaf native signing children in a series of studies, including naturalistic observation, lexical elicitation, fingerspelling, imitation of nonsense gestures, visual perspective-taking tasks, and a novel sign learning task. Shield hypothesised that an impairment in visual perspective-taking could lead to phonological errors in ASL, specifically in the parameters of palm orientation, movement, and location. Results showed that young deaf native signers with autism made frequent phonological errors involving palm orientation. These results suggest that deaf children with autism are impaired from an early age in a cognitive mechanism involved in the acquisition of sign language phonology.

Denmark compared deaf children with ASD and typically developing age- and language-matched deaf controls on a number of comprehension and production measures looking at emotional and linguistic use of the face in BSL. Deaf children with ASD performed comparably to deaf controls at comprehending and producing facial expressions across many of the tasks and showed no impairment with faces overall. Rather they showed specific difficulties with the comprehension and production of affective facial expressions and the comprehension and production of adverbial linguistic structures representing manner of action. These findings suggest that comprehension and production of affective and manner facial expressions in BSL rely on functions which are impaired in ASD, in contrast to other facial expressions which may be regarded as more purely linguistic. This provides a new line of evidence relating to separation of linguistic and other types of facial expression already described for signers. The lack of generalised impairments with faces in the deaf group in this study may also reflect the generalised advantage for all deaf children in face processing, related to the need to look to the face to communicate. Both studies demonstrate the importance of extending research on ASD to sign language users to enhance understanding of autism.

3.5. Tourette's syndrome

Gilles de la Tourette syndrome (TS) is characterized by vocal and motor tics starting in childhood. Vocal tics may be either noises or words, and vocal language tics may consist of obscenities (coprolalia) and repetitions of others' speech (echolalia). Sign language tics were first reported by Lang, Consky, and Sandor (1993) in a hearing woman with pre-existing TS who developed sign language tics following the learning of ASL as a therapeutic exercise in adulthood. Two later case studies described TS in a deaf adult (Morris et al. 2000) and a deaf child (Dalsgaard/Damm/Thomsen 2001), respectively. These two cases are of particular interest, since they provide evidence relevant to understanding the underlying cause of tics in TS generally.

The Morris et al. patient had the full array of tics seen in TS, but in sign language rather than in spoken language. The child "Sam", described by Dalsgaard, Damm, and Thomsen, produced sign language and spoken language tics. As with hearing TS patients, both produced obscenities as tics. These cases challenge several of the explanations advanced for the selective production of obscenities in TS. One of these is the idea that obscenities are produced by hearing people because of their expletive phonetic properties including harsh expiratory phonation. This explanation suggests that obscenities have different qualities than ordinary spoken words. It has also been sug-

gested that the production of obscenities may be a random process in which high-frequency phonemes are produced which happen to resemble obscenities. An alternative theory postulates a semantic origin: the meaning of obscenities may be the prime determinant of the tic rather than any underlying phonetic or phonological characteristic of the word itself, since the utterance of obscenities may reflect a failure to control brief aggressive impulses in TS. The obscene signs produced by these patients did not have any marked "expletive" quality in terms of amplitude of motion. Indeed, further evidence for a semantic basis for tics was provided by Morris et al.'s patient, who produced fingerspelled as well as signed obscenities.

4. Other studies

The various conditions described in this section differ from those in the previous sections since they are not directly concerned with either atypical development or with acquired impairments in a fluent user of a sign language. Instead they use cases of atypical individuals as testbeds for exploring neuroscience and linguistic theories. The first describes a study using the teaching of BSL as an L2 as a means of exploring the notion of the language module in relation to modality; the second expands this approach to studies of hearing individuals with developmental aphasia arising from epilepsy affecting the auditory cortex who have been taught sign language as an alternative to spoken language. In such cases – and unlike those discussed in the developmental section – there are clear dissociations between skills in spoken language and sign language. Studies of schizophrenic signers and stuttering provide evidence for differences in the nature of the feedback loops in spoken language and sign language.

4.1. A linguistic savant and sign language

Smith et al. (2010) report on an experiment in which Christopher (born 1962), a man with a remarkable ability for learning new languages alongside serious disabilities in other domains, was taught BSL. Christopher is mildly autistic and severely apraxic; he lives in sheltered accommodation because he is unable to look after himself. Perhaps uniquely, Christopher can read, write, speak, understand, and translate some 20 or more languages, while on tests of non-verbal intelligence he scores very poorly.

Christopher was exposed to a typical introductory course in BSL and his learning of BSL was compared to that of a control group of hearing university students. Christopher's general BSL learning was within the normal range of the control group's abilities (Smith et al. 2010). The one area where Christopher performed significantly worse than the control group was with comprehension and production of BSL Entity classifiers. In a task in which subjects had to match a signed sentence to a written English translation, Christopher scored 20 % correct; the scores of the control group were between 80 % and 100 %. In his processing of Entity classifiers, Christopher had some success identifying the class of referent that the handshape represented (curved versus straight objects, for example) but was not able to process the spatial location or movement that

the whole utterance encoded. Christopher did not go on to master BSL to a level comparable to that of his many other spoken second languages. Yet, he acquired an impressive single-sign lexicon both in comprehension and production and in doing so overcame his typical aversion to looking at people's faces when he communicates. His general BSL developed to a level comparable with other hearing sign language learners, but his acquisition differed from the control group in specific areas of the grammar. He was unable to overcome his difficulty with representing three-dimensional space and manipulations of these arrays in either the non-verbal domain or in linguistic mapping. His sign language abilities at the level of spatial syntax and morphology were thus limited by his cognitive impairments in non-verbal spatial processing. He did not experience this plateau in the acquisition of morphology in other second languages in the spoken modality and so his general cognitive impairment affects only his sign language learning. These results suggest fundamental modality-dependent differences for Christopher in the processing and learning of a sign language compared to a spoken language.

4.2. Landau-Kleffner syndrome

Landau-Kleffner syndrome (LKS) is an auditory agnosia which begins between the ages of 3 and 8 years and is thought to arise from an epileptic disorder within the auditory speech cortex. Typically, children with LKS initially develop normally but then lose comprehension and later production skills in spoken language. This loss is accompanied by an abnormal electroencephalogram (EEG) with the epileptic focus in the auditory cortex. The epilepsy usually subsides at puberty; however, a severe communication impairment often persists. Because of the intractable nature of the spoken language impairment associated with LKS, speech accompanied by signs has been used with this group, although this has largely not been successful (Bishop 1982). In contrast, several studies which have explored the acquisition and use of sign language in this population (Baynes et al. 1998; Sieratzki et al. 2001; Roulet Perez et al. 2001) have reported strikingly better sign language than spoken language

The case of Stewart (Sieratzki et al. 2001) illustrates this. His LKS began between 4 and 5 years, and he remains globally aphasic in English as an adult. He was initially educated in a school for children with severe language impairments, but because of a lack of improvement in his English language skills, he was transferred to a school for deaf children at the age of 13 years where he learned BSL.

Stewart demonstrates severe impairments in English on all measures with no improvement since childhood testing. At the age of 26, his scores for comprehension of grammar and vocabulary were equivalent to that of a 2 year old hearing child on a variety of standardised assessments. His performance on a picture naming vocabulary task in English was very poor, with only 17/50 responses correct in meaning and articulation. In contrast, in BSL, Stewart produced 29/50 items entirely correctly in meaning and articulation, and a further 13 items with single-parameter articulation errors. He was also assessed on BSL comprehension (Herman/Holmes/Woll 1999) and achieved a score corresponding to that of an average 9 year old native signer.

In individuals with LKS, the source of the impairment lies in processing of the auditory signal, with visual and spatial processing relatively unimpaired. The observa-

tions in section 2.1 above, that sign language comprehension may be more dependent on inferior parietal areas associated with somatosensory and visual motor integration while spoken language comprehension might rely more heavily on posterior temporal association regions whose input includes networks intimately involved with auditory speech processing, may underpin the relative sparing of sign language compared to spoken language for individuals like Stewart.

4.3. Stuttering

Dysfluencies in signing are known to occur in acquired neurological impairments such as Parkinson's disease, progressive supranuclear palsy, and motor neurone disease. Developmental dysfluency disorders in the deaf population are very rare, with a prevalence of stuttering in the deaf population of 0.4 % (Backus 1938) and 0.12 % (Montgomery/Fitch 1988), in contrast to the prevalence of stuttering in the hearing population of about 5 %. There have been a few surveys and case studies (Voelker/Voelker 1937; Backus 1938; Bloodstein 1969; Silverman/Silverman 1971; Montgomery/Fitch 1988). Interestingly, in many of these studies, sign stuttering is reported as most frequently occurring in simultaneous communication, where speech is co-articulated with sign.

Whitebread (2004) attempted to isolate and identify what might be the core features of stuttering in ASL. These potential features are: (1) inconsistent interruptions in sign and fingerspelling, (2) behaviours most often occurring at the beginning of the sign, (3) hesitation in sign movement, (4) repetition of sign movement, (5) exaggerated or prolonged signs, (6) unusual body movements unrelated to linguistic communication, (7) lack of fluidity in sign articulation, (8) inappropriate muscular tension associated with sign production, and (9) overt vocal or manual hesitation markers. The most recent study is Cosyns et al. (2009), a questionnaire study of stutter-like dysfluencies in Flemish Sign Language. Their respondents report observing 'involuntary interjections', 'repetition of sign movement', 'unusual body movements', and 'poor fluidity of the sign'. Strikingly, unlike speech stutterers, most signers exhibiting these behaviours were unaware of fluency problems. Additionally, dysfluencies were reported to occur at medial or final positions in signs, although in speech stuttering, repetitions or blocks at the end of a word are rare.

Modality-specific features of signing such as relatively less articulatory complexity, slower rate of articulation, etc. compared to speech − and of course a different set of articulators (see chapter 25, Language and Modality, for discussion) − may account for the strikingly low prevalence and atypical features of sign stuttering in comparison to speech stuttering, suggesting that stuttering may indeed be primarily a disorder of speech.

4.4. Schizophrenia and hallucinations of signers

The study of voice-hallucinations in signers (Schonauer et al. 1998; du Feu/McKenna 1999; Atkinson 2006; Atkinson et al. 2007) provides rare insight into the relationship

between sensory experience and how "voices" are perceived. The distinct way in which hallucinations are experienced may be due to differences in sensory feedback, which is influenced by both auditory deprivation and language modality. This highlights how the study of deaf people may inform wider understanding of auditory verbal hallucinations and subvocal processes generally.

There are crucial differences in how hallucinations are experienced by deaf and hearing people. Hearing people report vivid auditory imagery during voice hallucinations, whereas deaf people, although reporting voice hallucinations, are uncertain about auditory properties and often report visual or somatic analogues. It is possible that these differences may arise from differences in the perceptual feedback loop thought to be related to subvocalisation during thinking. Subvocalisation is primarily a form of motor imagery; perceptual feedback may vary depending on the modality of the subvocal articulation. Thus, a hearing individual might perceive an auditory trace ancillary to motor subvocalisation of their thoughts, and the same process may result in a visual or kinaesthetic percept for signers. Du Feu and McKenna (1999), for example, report on a deaf patient who perceived his thoughts as simultaneously signed outside his own head as if he could see them. It is possible he was experiencing imagery of the articulations underlying his sub-articulated thoughts. This leads to the question of whether these hallucinatory images should be considered to be visual or motor representations. Signers may combine visual and kinaesthetic percepts into a central representation in a way that is similar to integrated auditory-kinaesthetic representations in hearing people. However, an important difference exists for deaf people producing signs because the visual feedback received during self-production is substantially different from that received during comprehension. It is likely that mental representations of sign language are primarily kinaesthetic because muscle feedback dominates an individual's experience of his own productions. This is supported by Emmorey, Korpics, and Petronio's (2009) report on the apparent absence of a role for visual feedback provided by concurrent visual monitoring of hand movement in the lower periphery of vision during sign production. Because sign language production involves tracking the arms in space, hallucinations might result in premotor kinaesthetic traces that might be experienced as bizarre somatic sensations. Further research is needed to develop feedback models for sign language and to advance our understanding of how modality and sensory feedback shape the perceptual quality of experience.

5. Discussion: Issues of modality

At the beginning of this chapter, the question was asked whether language impairments reside in a specific modality and are thus linked to difficulties with auditory or visual-spatial processing, or are modality-independent deficits. It is possible that these two options are not mutually exclusive. In developmental impairments, specific perceptual processing or cognitive difficulties in the learner might interact with properties of the language modality. For example, difficulties in processing rapid sequences of closely related phonemes might create problems for the child acquiring a spoken language but might be less problematic for the acquisition of a sign language. Conversely, cognitive difficulties with representing three-dimensional space might not be crucial for acquir-

ing spoken languages but might prevent learners of sign languages from fully mastering the grammar. This may be the case with Heather (Williams syndrome) and Christopher (linguistic savant) although the impact of a visual-spatial impairment on each individual's sign acquisition was different. Heather, who learned to sign in childhood, was able to circumvent her visual-spatial problems with BSL and become a skilled signer, although with some abnormalities. Christopher, despite being a superlative language learner, found the morphosyntax-space interface very difficult to master, perhaps because he was exposed to BSL much later than Heather and used BSL far less (Smith et al. 2010).

A second possibility is that difficulties with language acquisition (whether signed or spoken) represent core processing problems at a higher level than those associated with the perception of the signal. A difficulty with the representation and processing of grammatical rules which allow the child to build up knowledge of the morphosyntactic regularities of the language being acquired would affect complex morphosyntax in both modalities, as appears to be the case with Ruthie and Sallie (Woll/Grove 1996; Woll/Grove/Kenchington 1998). The various cases discussed in this chapter provide contrasting evidence to address the question of whether different language impairments originate from cognitive, linguistic, or perceptual systems. In some cases, similar impairments are found in both modalities, suggesting an impairment independent of modality. In other cases, subjects show differences in language abilities in the two modalities.

In relation to acquired impairments, aphasia studies to date provide ample evidence for the importance of the left hemisphere in mediation of sign language in the deaf. Following left hemisphere damage, sign language performance breaks down in a linguistically significant manner. In addition, there is growing evidence for the role of the right hemisphere in aspects of sign language discourse and syntax. As well as their usefulness in illuminating the nature of aphasia breakdown, descriptions of sign language structure have raised new questions concerning the hemispheric specificity of linguistic processing. Two examples of specific interest identified by Corina are the case of sign paraphasia (patient RS) described by Hickok et al. (1996), and Atkinson et al.'s (2005) study of British signers with left- and right-hemisphere strokes, discussed in section 2.

These cases demonstrate the way in which a language's modality may uniquely influence the form of the linguistic deficit – in these cases, impairments with no clear parallels to spoken language disruption. The first study, the case of RS, is important for our understanding of the neurobiology of language, as the errors can be taken as evidence for selective language-form-specific linguistic impairment, indicating that the modality and/or form of a human linguistic system may place unique demands on the neural mediation and implementation of language.

The second study identified by Corina as of critical importance in addressing the relationship of modality and language is Atkinson and colleagues' (2005) study, which found that RHD signers, although generally exhibiting intact language, were impaired on tests of locative relationship expressed via classifier constructions and on classifier placement, orientation, and rotation.

Our understanding of the neural representation of human language has been greatly enriched by the consideration of sign languages of the deaf. Outwardly, this language form poses an interesting challenge for theories of cognitive and linguistic neural spe-

cialization, which classically have regarded the left hemisphere as being specialized for linguistic processing, and the right hemisphere as being specialized for visual-spatial abilities. Given the importance of putatively visual-spatial properties of sign forms (e.g., movement trajectories and paths through three-dimensional space, facial expressions, memory for abstract spatial locations, and orientation of the hands towards the body, etc.), one might expect a greater reliance of right hemisphere resources during sign language processing. However, despite major differences in the modalities of expression, striking parallels in the psycholinguistic and cognitive processing of these languages emerge (see Corina/Knapp (2008) and MacSweeney et al. (2008) for reviews).

6. Conclusion

The report of the UK government's Foresight Cognitive Systems Project (Marslen-Wilson 2003) identified the potentially unique contribution of sign language research to understanding how the brain processes language: "A more dramatic type of cross-linguistic contrast that may be uniquely valuable in elucidating the underlying properties of speech and language, comes through the comparison between spoken languages and native sign languages, such as BSL" (p. 9). As Corina suggests in relation to signers with stroke, such studies raise the question of whether, for example, aphasic deficits should be solely defined as those that have clear homologies to the aphasias arising from left hemisphere damage that are evidenced in spoken languages, or whether the existence of sign languages will force us to reconsider the conception of linguistic deficits.

The studies presented in this paper are examples of how cases of sign language impairments can provide a unique perspective and a model for investigating how different language impairments originate from different parts of the cognitive, linguistic, and perceptual systems. They also enable direct study of impairments in the context of cross-modal bilingualism. Finally, such profiles provide an evidence base for the development of appropriate interventions for use with deaf and hearing children.

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Bencie Woll, London (United Kingdom)