Short Title:
Language impairments in the development of sign
Full Title:
Language impairments in the development of sign: Do they reside in a specific modality or are they modality-independent deficits?

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Abstract

Various theories of developmental language impairments have sought to explain these impairments in modality-specific ways – for example, that the language deficits in SLI or Down syndrome arise from impairments in auditory processing. Studies of signers with language impairments, especially those who are bilingual in a spoken language as well as a sign language, provide a unique opportunity to contrast abilities across language in two modalities (cross-modal bilingualism). The aim of the paper is to examine what developmental sign language impairments can tell us about the relationship between language impairments and modality. A series of individual and small group studies are presented here illustrating language impairments in sign language users and cross-modal bilinguals, comprising Landau-Kleffner syndrome, Williams syndrome, Down syndrome, Autism and SLI. We conclude by suggesting how studies of sign language impairments can assist researchers to explore how different language impairments originate from different parts of the cognitive, linguistic and perceptual systems.

1. Introduction

Research into language acquisition has benefited greatly from a diversification of the populations and languages studied. Cross-linguistic comparisons of children developing language began to move the focus away from English to different language families and different language typologies (e.g. Slobin, 1985; Berman & Slobin, 1994). At the same time a new insight into the relationship between modality and language acquisition has been achieved by the study of children learning sign languages as native languages from their deaf parents (Newport & Meier, 1985; Petitto & Marentette, 1991; Anderson & Reilly 1998; Morgan & Woll, 2002). Further expansion of the field included studies of children growing up exposed to more than one language (e.g. Genesee, Nicoladis & Paradis, 1995). Research with children who display asynchronies in cognitive or language development can shed light on the complex developmental interactions between the linguistic and non-linguistic domains in typically developing children (Karmiloff-Smith, 1998). Additionally, our understanding of language acquisition in typically developing children has been enhanced by the documentation of language acquisition in populations where language and cognitive development is atypical because of conditions such as Down syndrome, Autistic Spectrum Disorder and Williams syndrome (e.g. Laws & Bishop, 2003; Tager-Flusberg, Plesa-Skwerer, Faja, & Joseph, 2003; Thomas, et al.., 2003). Interest in bilingualism and atypical development has been exemplified by studies of Specific Language Impairment (SLI) and Down syndrome in bilingual populations (e.g. Paradis, 2007; this volume; Kay-Raining Bird, Cleave, Trudeau, Thordadottir, & Sutton (2005).

The study of children exposed to a sign language but following an atypical course of development because of developmental disorders is an exciting new area of study. In this population, the questions are particularly complex: is language development affected by impairments in modality-specific or modality-independent ways? For example do children with sign language impairments have particular difficulty acquiring verb morphology or is this a consequence of SLI only in spoken languages? In cases where non-verbal cognitive deficits are present, how will these impact on the acquisition and use of a language perceived and produced in the visuo-spatial modality? For example, how does an impairment in processing non-linguistic visuo-spatial information impact on a child's acquisition of a signed language?

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 signed 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. In this paper we address the question of whether there are modality-independent language impairments which appear across both signed and spoken language acquisition, by reporting on a series of case studies of deaf and hearing individuals with cognitive or linguistic impairments which impact on their acquisition of British Sign Language (BSL).

1.1 Properties of the sign language signal and its processing

There are some important similarities and differences in the signal properties between signed and spoken languages (e.g. Brentari, 1998; Meier, 2002; Morgan, 2005) and these may be important for how different profiles of linguistic and cognitive impairments manifest themselves in learners of sign language.

Movements of the hands, arms, and body during signing are much larger than the movements of the articulators used for speech, and so the articulation of individual signs is about 1.5 times slower than for words (Emmorey, 2002). However, propositional rate is identical in sign and spoken language, as signers distribute grammatical devices across both hands and the face simultaneously, rather than in a more linear sequence as in spoken language (Bellugi & Fischer, 1972). The phonotactic structure of the sign signal has also some important properties which may influence how language impairment manifests itself. In particular, signs are largely mono or bi-syllabic and there are physical transitions in space between signs (e.g. Brentari, 1998; Orfanidou et al. 2010). In contrast speech is characterised as rapid sequences of phonemes without overt sound gaps between words (McQueen, 1998) and one difficulty children with SLI have is in efficiently segmenting the speech stream to identify multi-syllabic words boundaries. It follows then that the slower and less syllabically-heavy sign stream might not cause problems for SLI children exposed to BSL.

Beyond the word level, one way in which sign languages appear very different from spoken languages is that they exploit space for grammatical purposes. For example, grammatical markers of agreement appear on a discrete set of verbs in the lexicon that move between indexed locations in space. Agreement (co-location) links pronouns and noun phrases to their dependent referents and verb arguments, thereby indicating who did what to whom (see Sutton-Spence & Woll, 1999). Sign languages can also directly represent spatial relationships and physical forms. In a BSL sentence such as '*The man took down the hat from the top shelf and put it on his head'*, the shape of the hands in the verb represents handling a hat brim, the orientation of the hands represents the orientation of the hat, and the downward movement represents the actual path of movement of the action. Because structures of this type imply a direct mapping of real-world relationships in language, they have been described as utilising topographic space (Sutton-Spence & Woll, 1999).

Children with SLI acquiring spoken languages with rich verb morphology display patterns of errors different from children acquiring languages with limited verb morphology (Leonard, 2009). Sign languages also exhibit rich morphology through the presence of polymorphemic structures that resemble noun classifiers in spoken language (Supalla, 1986; Emmorey, 2003; Morgan & Woll, 2007). Entity classifiers in sign languages represent classes of nouns (e.g. flat entities, humans, animals, stick-like entities, etc.) and are essential components of spatial verbs (verbs of location and motion). The handshape encodes the class of entity and substitutes for the noun throughout the predicate (for more details, see Sutton-Spence & Woll, 1999). However, despite the striking differences in the surface forms of signed and spoken language, sign languages appear to be processed in the brain in largely similar ways to spoken languages (see MacSweeney et al. 2009).

2. Sign languages: acquisition

In order to understand how sign language development can be impaired we need first to document typical development of sign language in children. Children who are exposed to sign languages from early childhood show remarkable parallels in onset, rate, and patterns of development compared to children learning spoken languages (see Chamberlain, Morford, & Mayberry, 2000; Morgan & Woll, 2002; Schick, Marschark, & Spencer, 2004, for reviews).

Infants exposed to sign language from birth produce manual babbling at the same age as vocal babble emerges (Petitto & Marentette, 1991). The first 10 signs are produced around 12 months of age, and the 50 sign milestone is recorded from 20 months onward (Mayberry & Squires, 2006; Woolfe, Herman, Roy & Woll, 2010). Children combine signs from 18 to 24 months, initially using uninflected noun and verb forms (Newport & Meier, 1985; Morgan, Barrière, & Woll, 2006). Following the two-sign stage, children begin to produce more complex aspects of sign language grammar: articulating the location and movement of signs in space to express linguistic relations, and using a rich set of morphological markers (Supalla, 1986; Anderson & Reilly, 1998; Schick 1990; Morgan, Herman, Barriere, & Woll, 2008).

Bilingualism is common in children learning a sign language, since there is emphasis for deaf children on acquiring the spoken/written language of the majority community. The typical language learning environment for deaf children is by nature atypical. Fewer than 10% of deaf children have deaf parents who use sign language, and therefore few are native signers (Mayberry & Eichen, 1991). The vast majority of deaf children have an atypical amount and quality of exposure to spoken and signed language. Variability is found in age of first exposure; for example, spoken language may only be accessible following cochlear implantation; sign language may not be offered to a child until after failure to learn a spoken language. Variability is also found in quality of exposure: limited quality and amount of sign language input because of parents' limited sign language skills; limited quality and amount of spoken language input because impaired hearing limits access). Additionally, deaf and hearing children exposed to signed and spoken languages from birth onwards represent a unique type of bilingualism. Cross-modal (sign language and spoken language) bilingualism also presents a different context for language acquisition than unimodal bilingualism (two spoken languages or two sign languages), since the opportunities for code mixing in both input and output are different, with code-blending (the simultaneous articulation of a sign and a word) available as well as code-mixing (van den Bogaerde & Baker (2009) .

3. Hypotheses and predictions concerning modality and sign language impairment

The aim of this paper is to examine what two types of studies (hearing and deaf individuals with developmental impairments in sign language acquisition) can tell us about the relationship between language impairments and modality. In addressing the central question of whether language impairments reside in a specific modality, or are an outcome of modality-independent deficits, three alternative hypotheses can be formulated:

1. If the source of the impairment arises in processing of the auditory signal, with visual and spatial processing relatively unimpaired, no serious problems should be anticipated in the acquisition of sign language (performance in BSL is better than English).
2. If the source of the impairment arises in visual and spatial processing, and auditory perception is relatively unimpaired, no serious problems should be anticipated in the acquisition of spoken language but there may be deficits in sign language acquisition (performance in English is better than BSL).
3. If however, the problems relate to linguistic, as opposed to more general cognitive abilities, then delays and difficulties should be seen in language, regardless of modality (performance in sign language is similar to that reported for spoken language impairments).

In order to test these three hypotheses we review a series of case studies of atypical sign language acquisition and ask how the data under review provide evidence or counter-evidence for these predictions.

4. Case and group studies of language impairment in sign language

Two types of studies are presented here: in section 4.1 hearing children and young people who are atypical speakers of English and who also use BSL. These are Stewart, a hearing young man with Landau-Kleffner Syndrome, aphasic in English but with relatively good BSL; Christopher, the linguistic savant who learned BSL as an adult despite cognitive and language impairments; Ruthie and Sallie, identical hearing twins with Down Syndrome who are children of deaf parents; and in section 4.2 deaf children and young people who use BSL (and who also use English). These are Heather, a young deaf woman with specific visual-spatial impairments (Williams Syndrome); Paul, a case study of a deaf native signer of BSL; and finally, a group study of Deaf children with SLI in BSL.

4.1 Hearing users of sign language with atypical development

4.1.1 Stewart

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 language skills. This is accompanied by an abnormal electroencephalogram (EEG) with the epileptic focus in the auditory cortex. Although the EEG is always grossly abnormal, many children with LKS have no clinical seizures. The epilepsy usually subsides at puberty; a severe communication impairment often persists.

Stewart is a left-handed male who was a young adult at the time of the study (Sieratzki, Calvert, Brammer, Campbell, David & Woll, 2001). His LKS began between 4 and 5 years, and he is still globally aphasic in English. 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.

Performance in English

Stewart demonstrated severe impairments in English on all measures, including impaired phonological discrimination as measured on subtests of the Psycholinguistic Assessments of Language Processing in Aphasia - PALPA (Kay, Lesser & Coltheart, 1992) and poor syntactic ability, measured by the Test of Reception Of Grammar - TROG (Bishop, 2003). His performance on the TROG was extremely poor, characterised by errors with verbs, plurals, comparatives, passives and locatives. His scores from previous language testing were available and demonstrated no improvements since childhood (see Table 1 and Sieratzki et al.., 2001 for additional details).

[insert Table 1 here]

Stewart's reading ability was around the 7 year-old level on a variety of standardised tests. As part of the study undertaken when Stewart was 26, fifty items from the Snodgrass and

Vanderwart (1980) picture set were presented to Stewart who was asked to name the object in spoken English and then in BSL. In English, he produced 17/50 responses with correct meaning and articulation or only minor errors. He made phonological errors of an apraxic nature in over 50% of phonemes in 15/50 items and produced semantic errors on 8 items. Ten responses were unintelligible and uncategorisable in terms of semantic or phonological similarity.

Performance in BSL

A similar analysis was undertaken for Stewart's responses in BSL to the Snodgrass and Vanderwart (1980) set of pictures, using recognised articulatory parameters, *i.e.* combinations of handshape, location, movement, and hand orientation. Stewart produced 29/50 items entirely correctly in meaning and articulation, and a further 13 items with single-parameter articulation errors. Ten of the 13 errors were in sign movements, with a tendency to perseverate or enlarge movements, and there were 3 handshape errors. Only 2/50 responses showed dual-parameter errors in both movement and handshape. There were no errors in location or orientation. Non-articulatory errors occurred in 6/50 responses.

A BSL vocabulary comprehension test patterned after the British Picture Vocabulary Scales but designed to exclude iconic items which can be guessed by non-signers was administered. Although the test was not normed, mean age scores were available from a previous study with 70 deaf children age 4-11 years (Kirk et al., 1990). Stewart achieved a score of 54/68, exceeding the mean score of 45/68 for 11-year-old deaf children of hearing parents, and estimated to correspond to the expected performance of a 14-year-old. Eight of Stewart's 14 errors occurred in a single sequence close to the end of the test, raising the possibility of a transient absence.

Stewart was also assessed on a BSL grammar comprehension test standardised on native signing children aged 3-11 years (Herman, Holmes & Woll, 1999). Eleven year old native signers score near ceiling on this test, as the acquisition of BSL morphology is essentially complete by this age. Stewart scored 28/40, equivalent to an average 9 year old native signer, and corresponding to performance of a 12-year-old deaf child of hearing parents (see Table 1 above). Sign language, which Stewart first learned at the age of 13 years, is thus by far his most efficient communication modality. He has normal vocabulary and can process articulatory elements, implying the use of phonological mechanisms in BSL. He shows, however, strikingly uneven scores across the various subtests (number/distribution, negation, noun/verb distinctions, spatial verbs, size and shape specifiers, and handling classifiers), with high scores on spatial verbs and number/distribution but poor scores on negation, in contrast to lower scores across all subtests for deaf children of hearing parents. The uneven pattern Stewart exhibits is more typical of late learners of sign language as a primary language, *i.e.* subjects born deaf who are only exposed to a sign language after childhood, following failure to acquire a spoken language. However, having learned English early in life, Stewart does not fit straightforwardly into this category. It is of interest whether introduction to BSL earlier than age 13 would have enabled him to achieve a higher level of syntactic competence.

4.1. 2 Christopher

Christopher (born 1962) possesses a remarkable ability for learning new languages along side serious disabilities in other domains. He is mildly autistic and severely apraxic; he lives in sheltered accommodation because he is unable to look after himself..

Performance in English (and other spoken languages)

Christopher's knowledge of English syntax is essentially normal. Perhaps uniquely, he can read, write, speak, understand and translate some 20 or more languages while on tests of non-verbal intelligence he scores relatively poorly (see Smith and Tsimpli, 1995).

Performance in BSL

At the time of the study (Morgan, Smith, Tsimpli & Woll, 2007), Christopher was exposed to typical first course in BSL which ran for 8 months and included 24 hours of instruction in both taught and conversational modes. He also had access to BSL books and videos which he studied between classes. All his exposure to BSL came from native adult signers. We compared Christopher's learning with a control group of 40 (30 female, 10 male) hearing University students. They were taught in groups and were exposed to the same content (although over a shorter time period) as Christopher by a native BSL signer.. Christopher's general BSL learning was within the normal range of the control group's abilities (Smith, Tsimpli, Morgan & Woll, 2010). The one area where Christopher performed significantly worse than the control group, was with comprehension and production of BSL entity classifiers(see Section 1.1 above). In a task in which subjects had to match a signed sentence to a written English translation, Christopher scored 20% correct (chance was 33%); the scores of the control group were between 80% and 100% (mean 89%, SD = 9.9%). In a second task involving the matching of a signed sentence to a picture Christopher scored 10% correct (chance was 25%), whereas the controls scored between 50% and100% (mean 72%, SD = 13.8%). Compared with controls Christopher's performance on both tests was therefore extremely poor (see Morgan, et al. 2007 for more details of tests). In his processing of entity classifiers Christopher had some success identifying the class of referent that the handshape represents (curved versus straight objects for example) but was not able to process the spatial location or movement that the whole utterance encoded. For instance, after seeing the sign sequence CL-Bent-B-BOOK-ON-CL-B-BED which translated as 'a book on a bed', Christopher chose the picture of 'a book under a bed', rather than either of the other pictures which showed 'a ball under a chair' and 'a comb on a bed'. Christopher had particular difficulty with the classifier component of signing while performance of the controls was consistent across different BSL domains. These results suggest that there is a fundamental modality-dependent difference for Christopher in the processing and learning of a second language.

4.1.3 Ruthie and Sallie

Ruthie and Sallie are monozygotic twins who were born in May 1985 and have Mosaic Down syndrome (DS).. Both parents are deaf and members of the Deaf community. At the time of the study (Woll & Grove, 1996), the twins were 10 years old, and being educated in a mainstream setting, attached to a unit for children with special needs in their local primary school. In the presence of their parents and other deaf people they 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). They also produce occasional single-word English-only utterances and utterances produced with simultaneous sign and voice addressed to their parents.

In the presence of hearing children and adults and when playing together, they use English. They have not been observed to use BSL with each other when there are no deaf adults present. In this sense English appears to be their dominant language.

Assessments of the twins' verbal and nonverbal ability show that nonverbal cognitive skills are in advance of their verbal skills**.**

[insert Table 2 here

Performance in English

As can be seen in Table 2 above, Sallie's scores are consistently higher than Ruthie's, except for manual gesture. On comprehension measures of vocabulary (BPVS) and grammar (TROG), they are functioning between the level of three and four years. Both show evidence of developing morphology and simple syntax in a range of English structures, including negation, plurals, locative constructions, and interrogatives. Overall, the pattern of scores suggests that, as might be expected, visual and motor skills are relative strengths for both girls.

Performance in BSL

The twins show relatively higher skills for BSL vocabulary comprehension than for English. This pattern is even more apparent in a second round of data collection undertaken when the twins were 16 (Grove & Woll, in preparation) with BSL vocabulary continuing to increase while English vocabulary remained relatively static. It may be that this sign advantage is related to the presence of iconicity in many signs and the consequent resemblence of signs to gestures. Although typically developing children show no effect of iconicity in the acquisition of sign language, this may not be the case for atypically developing children. The lexical advantage for signs is not seen in morphology. Receptive skills were tested using the BSL Receptive Skills Test (Herman et al., 1999). Expressive skills were tested by asking the twins to describe pictures from the Receptive Skills Test. Sallie's BSL is more advanced than Ruthie's, but neither Sallie nor Ruthie has full mastery of the adult BSL system; 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 (English-like structures), rather than representing spatial relationships directly. Full details may be found in Woll & Grove, 1996).

Across various areas of morphosyntax in BSL, both girls have difficulty with those of the greatest complexity. These include structures requiring simultaneous marking of morphology and three-dimensional representations of space. In conclusion, Ruthie and Sallie apparently find the grammatical system of a sign language no easier to master than that of a spoken language.

4.2 Deaf users of sign language with atypical development

4.2.1 Heather

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 suggest that language in WS may be 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, 2007).

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. English speakers with WS show subtle linguistic impairments that may be related to problems with visuospatial cognition. These findings suggest either that language impairments may be arise from impairments in visuospatial cognitive domains or that spatial aspects of both cognition and language are controlled by a higher-level representational system.

One group of languages for which the consequence of specific visuospatial learning difficulties might be particularly severe is sign language. 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.

Heather is a young deaf woman aged 34 years at the time of the study (Atkinson, Woll & Gathercole, 2002). 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 was educated in a school for children with learning disabilities where the Makaton and Paget-Gorman sign systems were used, together with rudimentary BSL. Heather first came into contact with adult Deaf native signers at 14 years of age. Little is known about the quality of her language models prior to this age. 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 immediately apparent in spontaneous conversation, she does make consistent errors in her use of some features of BSL. The precise nature of Heather's visuospatial difficulties were investigated using standardised tests of visual and spatial abilities (see Table 3).

[insert table 3 here]

These results show clear impairments in visuospatial ability, in contrast to Heather's preserved ability to discriminate faces: she scored 48/54 on the Benton Test of Facial Recognition (Benton, Hamsher, Varney, & Sprren, 1983), which is in the middle of the normal range for adults. It is clear that Heather shows the dissociation between intact face recognition and impaired visuospatial construction which is characteristic of the WS profile.

Performance in BSL

Heather was tested both on comprehension and production of spatialized syntax at sentential level on connected discourse. The BSL production test (Herman, et al.. 2004) although designed for use with children, allowed Heather to be assessed on BSL narrative in a systematic way. This test assesses deaf signer's expressive language by eliciting a narrative. The participant watches a short language-free story acted out by two deaf children, which is presented on a DVD. The participant is then asked to tell the story, which is video-recorded for subsequent scoring. The assessment is scored in three parts: (1) the prepositional content of the story (i.e. how much information children include in their narrative), (2) structural components of the narrative (i.e. introducing the participants and the setting, reporting the key events leading up to the climax of the story, and telling how the story ends) and (3) aspects of BSL grammar (including use of spatial location, person and object classifiers and role shift.

Heather displayed marked problems with structures using space for grammatical purposes. At sentential level these difficulties include problems with ensuring that verbs correctly indicate semantic roles. Heather's production of spatial verbs shows consistent impairment in spatial representations. 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. Static locatives using topographic space are rarely used (e.g. *the pencil is on the table* should be produced by first signing TABLE, then PENCIL, and then placing the classifier for PENCIL in a location in space immediately above where TABLE was signed. Instead, Heather prefers signing PENCIL ON TABLE, without the required spatial relationship between the signs.) 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).

In the context of the narrative, Heather also had 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 provides an interesting comparison with Christopher in this regard.

4.2.2 Paul

Paul is a congenitally deaf son of deaf parents, aged 5;2 at the time of the study (Morgan, Herman, & Woll, 2007). Paul was exposed to fluent BSL from birth and from 2 years attended a mainstream kindergarten and later a school with sign language support. He was referred for assessment by the school because of worries about his BSL development which was described as being unusually slow for a native signer. His non-verbal cognitive abilities, assessed with the Snijders-Oomen nonverbal intelligence test (Snijders, Tellegen & Laros, 1989) when he was 5;0, revealed no cognitive delays. It might be expected that SLI in BSL would affect comprehension and production of structures involving polymorphemic verbs (agreeing, spatial and classifiers) and complex syntactic structures involving simultaneous manual and nonmanual markers (negation). His BSL grammar was assessed using the BSL Receptive Skills Test (Herman et al., 1999). Paul scored 1.3 standard deviations below the mean for grammar. His performance was atypical, with success on some difficult items, and failure on many easier ones. He was particularly poor on negation, spatial verbs and classifiers.

We elicited production data of BSL grammatical structures by asking Paul to describe pictures taken from the BSL receptive skills test (Herman, et al., 1999) . For example Paul was asked to describe a picture of a man giving a boy a letter. Typically developing native signers of his age use inflectional morphology on the verb GIVE to indicate subject and object: MAN LETTER GIVE-3 (*the man gives the letter to him/her*). In contrast Paul signed the following sequence of uninflected signs (P=Paul A= Deaf adult).

P: GIVE GIVE SQUARE GIVE (citation forms) *give, give the square thing give*

A: SQUARE GIVE WHO?

who gives the square thing?

P: GIVE GIVE POINT (picture) LETTER

give, give, (point), letter

A: PICTURE WHAT?

what is in the picture?

P: LETTER POINT (PICTURE)

a letter (point)

Paul was also tested on a pilot version of the Non-Sign Repetition Test (Marshall, Denmark & Morgan, 2006). This assessment tool evaluates the participant's ability to copy a set of 40 nonsense but possible BSL signs of varying phonological complexity (for completed test and norms see Mann et al., 2010). The pilot version was administered to Paul and a group of 18 native signers aged between 2 and 10 years (see figure 1). Paul's performance was severely impaired (below the score of a 2y;6m control).

[insert figure 1 here]

Nonword repetition has been reported to be a robust marker of SLI in children acquiring spoken languages. However Stokes, Wong, Fletcher and Leonard (2006) tested Cantonese children using multi-syllabic non-words and found no difference between SLI and age-matched control children. The authors suggested that this was because Cantonese does not contain the complex phonotactic structures, variable stress patterns, and difficult-to-articulate consonants that make non-word repetition in languages such as English and Swedish so difficult.

From this case study it was hypothesized that SLI in sign language would affect those areas previously identified as fragile in spoken language acquisition – specifically complex morphological marking and non-word repetition. In order to confirm this hypothesis a group study was undertaken..

4.2.3 SLI group study.

The SLI group study is of 13 deaf children aged 5;10 to 14;8 whose first language was BSL. The children were referred for assessment by teachers or speech and language therapists because of concerns about their sign language development in comparison with their peers (Mason et al., 2010). The youngest child (aged 5;10) had deaf parents; the remaining 12 children were aged between 7 and 14 years. Children under 7 years from hearing families were not included in the study so as minimise the possible effects of late exposure to sign language. All children had been exposed for at least 4 years to native signers of BSL. All had normal motor and cognitive development, with motor skills assessed using a bead threading task for which scores had been obtained for typically developing deaf children (Mann, et al.., 2010). Non-verbal cognitive abilities were assessed using three sub-tests of the British Ability Scales: pattern construction, matrices and recall of designs (Elliot, Smith & McCulloch, 1996). The children were also assessed on non-sign repetition, BSL receptive grammar and grammatical and pragmatic skills (Herman et al., 1999; Herman, Grove, Holmes, Morgan, Sutherland, & Woll, 2004). Table 4 presents the findings from these assessments. A shaded square represents performance considered impaired following standard criteria.

[Table 4 here]

To summarize these data, 7/13 children displayed impaired receptive grammar, 8/13 had impaired productive grammar, but only 4/13 had impaired non-sign repetition. More analysis of how typically developing children perform on this task is required before we discount sign phonology as a sign SLI marker (Marshall, Denmark & Morgan, 2006). However it was clearer that complex morphology did appear to be impaired in this group study. The results also suggest different profiles of impairment in individual children: phonological, receptive grammar, productive grammar, pragmatics and discourse (as measured by narrative structure). These findings suggest SLI appears to affect language acquisition in similar ways across modality but with language typology (sign languages do not have multi-syllabic word structure but do have complex verb morphology) also influencing which aspects of linguistic structure are more or less affected.

5. Discussion

In the questions posed at the beginning of this paper, we asked whether language impairments reside in a specific modality and are thus linked to acquisition difficulties with auditory or visual signals, or are modality-independent deficits. It is possible that these two options are not mutually exclusive. 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 may create problems for the child acquiring spoken language but may be less problematic for the acquisition of a signed language. Conversely, cognitive difficulties with representing three dimensional space might not be crucial for acquiring spoken languages but might prevent learners of signed languages from fully mastering the grammar. This may be the case with Heather and Christopher although the impact of a visuo-spatial impairment on each individual's sign acquisition was different. Heather being deaf and immersed in the deaf community was able to circumvent her particular problems with BSL and became a skilled signer. Christopher, despite being a superlative language learner found the morpho-syntax- space interface very difficult to master, perhaps because of his age of acquisition being later than Heather and the fact that he was hearing meant he used BSL far less than Heather (Smith et al., 2010).

The second option is that there may be difficulties with language acquisition (whether signed or spoken) that represent core processing problems at a higher level than those associated with the perceptual carrier of the signal. A difficulty with the representation and processing of grammatical rules which allow the child to build up knowledge of the morpho-syntactic regularities of the language they are acquiring would affect complex morpho-syntax in both modalities, as appeared to be the case with Paul (Morgan et al., 2007) and some of the children in the group study of SLI (Mason et al., 2010).

The various cases presented here 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 the sections below, we review our initial hypotheses in the light of data from these studies.

5.1 ***Hearing users of sign language with atypical development***

5.1.1 Spared BSL relative to English (Stewart)

The first hypothesis we considered predicted that if the source of the impairment arises in processing of the auditory signal, with visual and spatial processing relatively unimpaired, no serious problems should be anticipated in the acquisition of sign language. LKS is an auditory phonological processing disorder. Stewart's BSL is significantly better than his English despite very late exposure. Sign language thus appears to be an effective means of communication even in the face of severe spoken language aphasia for this group.

5.1.2 English = BSL (Ruthie and Sallie)

Where problems relate to linguistic, as opposed to more general cognitive abilities, it was hypothesized that delays and difficulties should be seen in language, regardless of modality. The twins' BSL grammar is at a comparable level to their English grammar. Sallie, the more able twin in English, is also better at BSL. This suggests a cross-modal linguistic deficit, with the pattern of varying competences in both languages related to the complexity of the required linguistic devices, and consistent with observations of difficulties in the acquisition and generalisation of rules affecting complex sentence structure in children with DS. Some studies of children with DS suggest 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, Mangan, Obrzut, & Nadel, 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. 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.

5.1.3 ***English > BSL (Christopher)***

If the source of the impairment arises in visual and spatial processing, and auditory perception is relatively unimpaired, it was hypothesized that no serious problems should be anticipated in the acquisition of spoken language but there might be deficits in sign language acquisition. Christopher was an adult learner of BSL but he did not go onto master the language as he has done for his many other spoken second languages. He did acquire 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 the non verbal domain in order to use physical space for 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.

5.2 Deaf users of sign language with atypical development

Evidence for three different patterns of impairment was also found in the deaf signers.

5.2.1 Impaired sign language relative to that reported for spoken English hearing individuals with Williams syndrome (Heather)

Heather's language abilities in general are well in advance of her visuospatial abilities. Her language profile differs from that of a hearing individual with Williams syndrome, with the subtle impairments that have been found in spoken language in Williams syndrome are more transparent in BSL. Most strikingly, 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 visuospatial language is not in itself dependent on intact visuospatial cognition (see Morgan, Smith, Tsimpli & Woll, 2002), 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, visuospatial impairment overrides general grammatical ability and Heather prefers to use English-like constructions which make fewer direct demands on visual spatial cognition.

5.2.2 Performance in the sign language modality is similar to that reported for spoken language impairments. (Paul and SLI group study)

The deaf children with SLI show comparable impairments to those found in hearing children with SLI. In both of the SLI studies, impairment was reported for sign language grammatical constructions involving verb agreement. Paul used uninflected verb forms despite being prompted by the deaf native-signer tester that these utterances were unclear. His difficulty in using BSL verb morphology might be linked to the nature of meaning-form mappings using agreement verbs in BSL. In changing the morphology of the verb by inflecting movement between two locations in sign space, signers map out the core meaning e.g. ‘giving’ but also the direction of the inflection simultaneously encodes argument structure (i.e. the identity of the agent and patient). This packaging of information into a single unit with several components requires good language skills. Paul preferred to map out each part of the proposition in a sequence of signs using points to agent and patient arguments and an uninflected verb. This type of error resembles the BSL produced by much younger typically developing signers (Morgan, Barriere & Woll, 2006). In the assessment of the production of BSL grammatical devices in narratives from the group study of sign SLI, 8 of the 13 children were impaired. The sets of data from Paul and from the group study suggest that SLI affects BSL verb morphology in similar ways in spoken and signed language (Leonard, 2009). Where this is the case, the inclusion of sign languages in research 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, 1984) or a domain specific linguistic impairment (van der Lely, 2005) but do suggest that modality-related processing difficulties cannot be the source.

6. Conclusions

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). 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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Table 1 – Spoken language and literacy from age 5;8 to 21 years

Age at Testing	Skill	Test	Standardised Age Equivalent
5;8	English language (RDLS[1])	Verbal comprehension	2y
13	English language (RDLS[1])	Expressive language Verbal comprehension	3y1m 2y2m
21	English language (BPVS[2])	Picture vocabulary (long form) Receptive vocabulary	2y4m 2y4m-3y
21	Literacy	BAS[3] Word Reading BAS[3] Word Spelling Neale reading[4]: accuracy Neale reading[4]: comprehension	7y6m 8y >7y 7y

[1]Reynell Developmental Language Scales; [2]British Picture Vocabulary Scales; [3]British Ability Scales; Neale Analysis of Reading Ability (Neale, 1997).

Table 2 age equivalents of Ruthie and Sallie's Test Results (age 10 years)

Tests (English and NV IQ)	Sallie	Ruthie
Snijders-Oomen (nonverbal IQ)	5;8	5;3
British Picture Vocabulary Scales (receptive vocabulary)	3;7	3;1
Test of the Reception Of Grammar	4;0	<4;0
Edinburgh Articulation Test (speech articulation)	5;6	4;0
Illinois Test of Psycholinguistic Abilities auditory memory	3;0	2;5
visual memory	4;4	3;7
manual expression	4;10	5;6
Tests (BSL)		
Receptive vocabulary	5;3	5;8
Receptive grammar	4;4	3;8

Table 3. Heather's performance on tests of visual and spatial abilities

DAS Pattern construction (Jarrold et al.. 1999 mean for WS adults = 8;4)	7;4
DAS Copying	6;10
Raven's Matrices (Raven, Raven & Court, 1998).	8;4
Visual Form Discrimination (normal range 24-30) Errors suggest difficulties with breaking images down into their constituent parts	17
BORB Orientation Match (normal range 24-30)	17
Benton Facial Recognition Test (normal range 41-54)	48
DAS Digit span (Signed with lip-pattern) equivalent to 4;4 (1st percentile at 17;6 –17;11 years)	3
Corsi Span	2

Figure 1.

Paul's performance (circled) on the Non-Sign Repetition Test compared to 18 CHECK native signers aged 2-10.

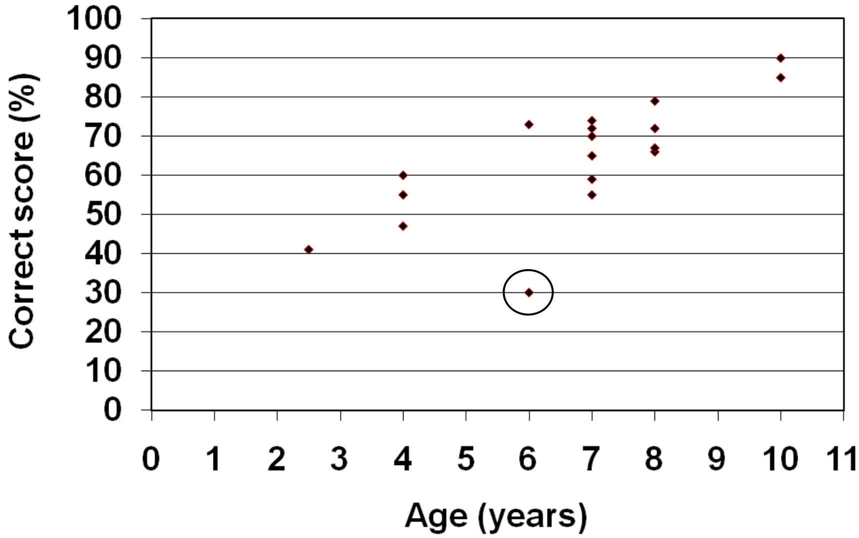

Table 4. Data for individual children with suspected SLI in BSL (from Mason et al., 2010)

Child	Age	BAS z-score	BSL Receptive Test z-score	BSL Production Test percentile scores			Non-Sign Repetition Test z-scores
				Narrative Content	Narrative Structure	BSL Grammar	
1	13;11	-0.6	0.3*	25*	50*	10*	0.6*
2	7;04	-0.6	<-2.1	<10	<10	<10	-1.3
3	14;02	-0.1	1.1*	10*	10*	25*	0.5*
4	14;08	-0.9	-1.5	10*	<10	10*	-0.1*
5	7;04	0.6	-2.1	<10	10	<10	1.1
6	11;0	-0.7	0.1	25	10	50	-1.7
7	5;10	-1.2	<-2.1	<10	10	25	0.7
8	8;01	-1.2	0.6	<10	<10	25	-2.0
9	9;01	-0.6	-2.3	10	25	10	0.9
10	10;06	.03	-1.5	<10	<10	<10	0.2
11	10;09	-0.5	<-2.1	<10	<10	<10	-1.4
12	9;08	0.7	1.1	<25	10	<25	-0.5
13	11;03	-1.0	-0.7	10	50	10	-0.3
Range	5;10 – 14;08	-1.2 – 0.6	-2.1 – 1.1	<10 - 25	<10 - 50	<10 - 50	-2.0 – 1.1

* child older than range for standardised scores