NEUROCASE
2009, 15 (5), 419–426

Psychology Press
Taylor & Francis Group

The effects of cerebellar ataxia on sign language production: A case study

Martha E. Tyrone,[1] Joanna R. Atkinson,[2] Jane Marshall,[3] and Bencie Woll[2]

[1]Haskins Laboratories, New Haven, CT, USA
[2]University College London, Deafness, Cognition, and Language Research Centre, London, UK
[3]Department of Language and Communication Science, City University London, London, UK

Speech and sign production both require precise coordination of multiple articulators. The characteristics of dysarthria following ataxia have been well-documented, but less is known about the consequences of ataxia for sign language, which uses the hands and arms as articulators. This is the first study to examine ataxic dysarthria in a sign language user. What is novel in this research is that the limbs are employed for both linguistic and non-linguistic movements. Notably, sign production deficits broadly resembled ataxic dysarthria, while non-linguistic movement deficits were similar to those previously reported for ataxic limb movement.

Keywords: Sign language; Dysarthria; Limb movement; Ataxia; Cerebellum.

BACKGROUND

Sign languages are the natural languages of Deaf[1] communities. They exhibit the same grammatical and semantic complexity as spoken languages but do not share the grammar and lexicon of the spoken languages that surround them. One basic difference between sign languages and spoken languages is that sign languages use the hands and arms, rather than the vocal tract, as primary articulators. Many of the resulting structural differences between sign and speech have been documented (Meier, 2002) but there has been little research exploring which of these differences are specifically articulatory, rather than perceptual or linguistic. The examination of sign production in a movement disorder such as cerebellar ataxia could help to address this question. This article reports on a British Sign Language (BSL) user with acquired cerebellar ataxia, who was part of a larger study on sign language and movement disorders (Tyrone & Woll, 2008a, 2008b).

Ataxia refers to the set of motor symptoms resulting from cerebellar damage, which typically include incoordination and movement inaccuracy (Timmann, Citron, Watts, & Hore, 2001), dysrhythmia, dysdiadochokinesia (disruption to rapidly alternating movements), intention tremor, and dysmetria (movement undershoot or overshoot) (Bastian, 2002; Topka, Konczak, Schneider, Boose, & Dichgans,

[1]This paper uses 'deaf' to describe hearing loss, and 'Deaf' to describe the cultural group of sign language users.

This work was funded by the Wellcome Foundation (grant 053239), the Deafness Cognition and Language Research Centre (Economic and Social Research Council UK grant RES-620-28-6001), a grant from the National Institutes of Health (DC6523), and an ORS studentship from the British Council. The authors would like to thank Trudi Collier, the sign model, and the two Deaf individuals who participated in the study.

Address correspondence to Dr Martha E. Tyrone, Haskins Laboratories, 300 George St., Suite 900, New Haven, CT 06511, USA (E-mail: tyrone@haskins.yale.edu).

http://www.psypress.com/neurocase
DOI: 10.1080/13554790902893097

1998). These deficits would have a profound impact on one's ability to sign, because sign production requires precise coordination of movements of the shoulder, elbow, wrist and fingers, both across limbs and within a single limb. Hand and limb movements must also be coordinated with movements of the torso and head. Moreover, many signs require that the hand change its configuration while the arm is in motion. In these cases, a signer must integrate proximal and distal movements (e.g., of the elbow and fingers) within a single sign. There has been no research on the consequences of ataxia for sign language, but given what is known about the types of movements required for fluent signing, ataxia is likely to severely disrupt sign production.

Ataxic speech is described as slow, distorted, and imprecise, with irregular variations in pitch and loudness and a 'scanning' rhythm (Kent et al., 2000). Clinical and experimental research indicate that ataxic dysarthria affects many speech articulators at once, rather than individual articulators in isolation (Kent, Kent, Rosenbek, Vorperian, & Weismer, 1997; Kent et al., 2000; Saigusa et al., 2005). In their comparison of ataxic speech and ataxic limb movements, Ackermann, Hertrich, Daum, Scharf, and Spieker (1997) emphasized the effector-specific differences between the two types of movement, arguing that dysmetria occurs for the latter but not the former. This study seeks in part to disambiguate the effects of movement task and effector by examining linguistic and non-linguistic movements of the hands and arms, which can be compared to findings from previous sign and speech research.

CASE STUDY

Robert[2] is a 36-year-old Deaf man whose movement deficits began 3 years prior to this study. Ataxia was caused by a large cerebellar infarct resulting from extensive hemorrhaging during surgery to correct an arteriovenous malformation. A second operation was performed to address the hemorrhage, and CT scan data were collected after the second operation during the same hospital stay. The scan showed damage to the right cerebellar hemisphere, the vermis and a small area of the pons. Since his operation, Robert has been unable to walk and uses a wheelchair. His posture is unstable, and he has irregular postural sway. His saccadic eye movements and his limb movements are severely disrupted and he performs self-care routines with difficulty.

Robert's primary mode of communication is BSL, although he has some speech and lipreading abilities. His family is hearing, and he communicates with them through lipreading and spoken English. His friends and peers are mostly Deaf, and his communication with them is in BSL. Since surgery, Robert's signing has been severely disrupted, with a strong impact on his intelligibility and consequences for his communication with others. As with most cases of ataxia, the production difficulties he exhibits are motoric rather than linguistic in nature. This is indicated by his ease in understanding everyday signed conversations and by his ceiling scores on tests of naming and comprehension of single signs and sentences in BSL (Atkinson, Marshall, Woll, & Thacker, 2005).

Signing task

Method

Robert was asked to perform a series of linguistic and non-linguistic motor tasks, including a signing task, in which he copied individual signs after they were produced by the experimenter. His performance was directly compared to the performance of a healthy Deaf control signer, who was matched for age and linguistic background. The two signers were each videotaped as they performed the linguistic and non-linguistic tasks, and all analyses of the tasks were carried out after testing. Sign productions were coded for the major sign parameters (handshape, location, movement, and orientation, the meaningless structural units that differentiate one sign from another, similar to phonemes in spoken language) (Battison, Markowicz, & Woodward, 1975; Stokoe, 1960) and for disruptions to non-linguistic aspects of sign movements (e.g., involuntary movement, or incoordination across articulators). To simplify the comparison across tasks, the parameters that Stokoe (1960) identified as 'location' and 'movement' are referred to here as 'targeting' and 'trajectory', respectively. The values of each of the sign parameters were coded, and deviations from the citation form of a sign were recorded for each sign that was produced by each participant. All the data were coded by the first author, and half the sign data were double-coded by a graduate student with training in sign language research and in speech and language therapy. The two coders had a 96% agreement rate.

[2]Not his real name.

Assessments of involuntary movements were based on perceptual measures and compared to reports from the literature (e.g., Brooks 1986; Fahn, Greene, Ford, & Bressman, 1998). The atypical signer described here only exhibited one type of involuntary movement, which was intention tremor. Given the exploratory nature of the study, descriptive measures were used for both the linguistic and non-linguistic tasks.

A description of the coding scheme is included in Table 1. Stimuli were selected to represent a range of sign forms and were balanced to include signs with and without handshape change, orientation change, and bimanual activity. Omission of a required change and addition of an extraneous change (e.g., handshape change) were both counted as a deviation from a sign's citation form; therefore, each category in the coding scheme was applicable to each sign. Thirty productions were analyzed for each signer.

A movement was coded as having a targeting error if there was either overshoot or undershoot of the movement target. Similarly, when a movement noticeably veered away from the direction of the movement target, it was coded as a trajectory error. Trajectory errors were distinguished from instances of intention tremor based on cyclicity and movement size. Small, cyclic movements that co-occurred with a targeted limb movement were considered tremors, while larger, non-cyclic deviations away from the direction of the movement target were considered trajectory errors.

TABLE 1
Sign coding scheme

Handshape: Anomalies in the internal configuration(s) of the hand(s) during sign production
Handshape: Change: Anomalies in changing or maintaining the handshape required by a sign
Orientation: Anomalies in direction of orientation(s) of the palm(s)
Orientation Change: Anomalies in changing or maintaining the palm orientation required by a sign
Targeting: Movement of the hand(s) to an anomalous location on the body or in space
Trajectory: Anomalies in the direction, number, and manner of elbow and/or shoulder movements
Self-repetition: Spontaneous repetition of one's own production of an entire sign without pause
Involuntary movement: Small, cyclic movements that co-occurred with targeted limb movement
Unimanual coordination: Incoordination of multiple articulators on one limb
Bimanual coordination: Incoordination of movements of the two limbs

Sign: results

In the course of normal production, signs, like spoken words, often deviate from their citation forms; thus, there are multiple 'anomalous' productions for both Robert and the control signer. Robert's productions deviated from citation form often and in ways that differed from the control's productions (Figure 1). First of all, intention tremor (i.e., involuntary movements that occur only during deliberate movement) was present in 18 out of 30 of his sign productions. There was no clear pattern in the distribution of intention tremor in his signing, and no involuntary movements were observed for the control. Robert also hyperextended his fingers during signing, resulting in anomalous handshapes for the target signs. Furthermore, he showed incoordination of the movements of proximal and distal articulators (e.g., the elbow and fingers) during signing. By contrast, the control's productions included few atypical handshapes and little proximal/distal incoordination. Moreover, the type of handshape errors made by the two signers were distinct. When the control's handshapes deviated from citation form, his fingers were laxed or hypoextended (Figure 2), whereas Robert produced hyperextended handshapes, in which the fingers were extended out from the hand such that they began to point dorsally along the arm (Figure 3).

Robert's motor symptoms often affected the dynamic components of signs, i.e., the elements that change configuration over the course of a sign's production. He had difficulty coordinating movements of articulators on the same arm (e.g., the elbow and fingers); and in some cases, he added movements (e.g., handshape change) to signs in which they were not required. Robert also had difficulty with handshape change and orientation change. Handshape change and orientation change were coded as not matching the citation form either when a configuration change that was required for correct production of a sign was omitted, or when a configuration change that was not part of the sign was added. By contrast, none of the control's sign productions included an anomalous handshape change, although a few included an anomalous orientation change: in two cases an extraneous change was added, and in one case a required change was omitted.

Similarly, Robert had difficulty with spatial and temporal coordination of the two limbs on two-handed signs; WITH (Figure 4) is an example

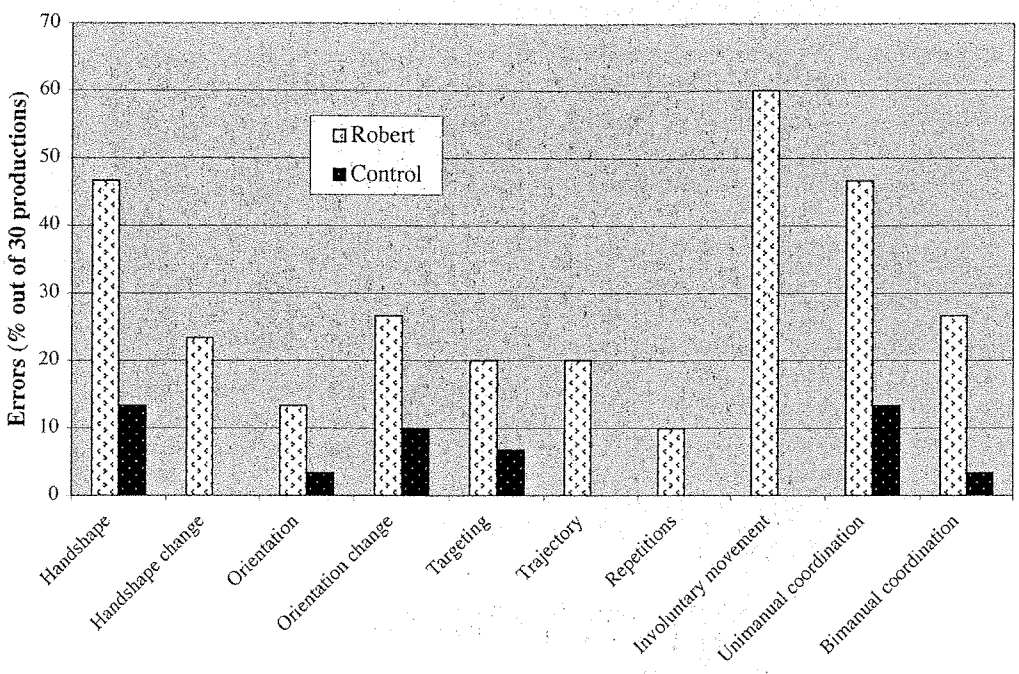

Figure 1. Sign Copying Task.

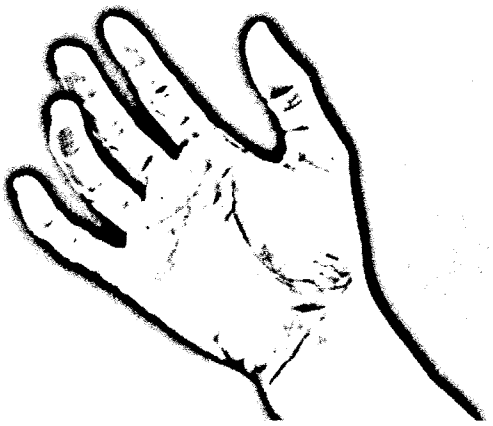

Figure 2. Laxed handshape.

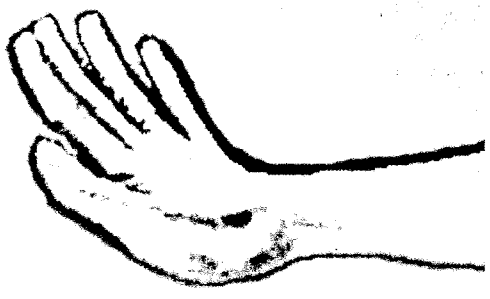

Figure 3. Hyperextended handshape.

Figure 4. BSL sign WITH.

of a sign that requires handshape change and coordination of the two limbs. On this sign in particular, Robert's two hands changed configuration at different times, and he repeated the hand-internal movement unnecessarily. In addition, he treated some one-handed signs as two-handed, by producing the same actions with his dominant limb and his non-dominant limb simultaneously. These were all counted as bimanual incoordination, which occurred in 26.67% of his productions.

Regarding static elements of sign structure, Robert tended to use articulators proximal to those normally used for a given sign (e.g., flexing the wrist instead of the metacarpal finger joints).

In addition, disruptions to fine motor control affected how he formed sign handshapes, such that his fingers and wrist were hyperextended.

Non-linguistic tasks

Method

Robert and the control participant were tested on several non-linguistic motor tasks. The two discussed here are a pointing task and a reach and grasp task. For the pointing task, the two participants were asked to point to one of two illustrations in response to a signed utterance produced by the experimenter. A response sheet with two illustrations on A4 size paper was placed on a flat surface directly in front of each participant. Incorrect responses and hesitations were excluded from analysis, in order to avoid motoric artefacts of a lexical search process. A total of 28 responses on the pointing task were analyzed from each testing session.

For the reach and grasp task, four cylinders of different diameters (4.5, 5, 6.3, and 8.5 cm.) were placed on a table in front of the participants. They were each asked to grasp the cylinders individually, move them approximately 25 cm forward, set them down, then move them back to their original locations. In total, 12 reach and grasp trials were analyzed from each testing session. The pointing task and the reach and grasp task were both coded according to the scheme in Table 2. The data collection and coding procedures are the same as those outlined for the signing task.

In the pointing task, Robert's proximal and distal joint movements were incoordinated on multiple trials, even though the task required no change in hand configuration from one production to another (Figure 5). Proximal/distal incoordination most often occurred as instances in which he moved his hand toward the target, hesitated, configured his hand for a pointing gesture, and then continued moving to the target. He formed the hand configuration again for almost every trial rather than maintaining it across trials, and the result was that he had difficulty coordinating the movements of multiple effectors simultaneously.

Dysmetria (i.e., targeting errors) occurred in 25% of Robert's pointing movements (Figure 5). Specific patterns in targeting errors were not quantified; however, in most cases, he overshot the target and moved his hand too far. Other times, he divided the individual movements into two components by moving his hand to the sheet of paper and then sliding it to the target. The control produced only one unexplained pause and one targeting error.

In the reach and grasp task, Robert had many targeting and coordination errors (Figure 6). Also, his hand configurations were often not well-adapted to the size and shape of the cylinders, and when this occurred it usually impeded his completion of the grasp. Additionally, he had multiple involuntary movements when performing the task. While he was given no explicit instruction on which hand(s) to use, Robert repeatedly used both hands for individual trials on the task.

TABLE 2
Non-linguistic coding scheme

Handshape: Atypical configuration of the hand for a particular movement task
Targeting: Inaccurate placement of hand relative to movement target
Involuntary movement: Small, cyclic movements that co-occurred with targeted limb movement
Pause/Hesitation: Cessation of movement during execution of a task
Speed: Unusually slow or rapid movement
Unimanual coordination: Incoordination of movements of proximal and distal effectors

DISCUSSION

It seems that aspects of signing which are most markedly impaired in Robert's case are related to the motor symptoms most common in ataxia: tremor during deliberate movement, incoordination of multi-joint movements (Topka et al., 1998; Tracy et al., 2001), and disruptions to fine motor control (Timmann et al., 2001). This is consistent with findings from earlier research on precision grip and reach and grasp tasks in cerebellar patients (Fellows, Ernst, Schwarz, Topper, & Noth, 2001; Zackowski, Thach, & Bastian, 2002). Bastian (2002) proposes that the cerebellum plays a specific role in motor control by adjusting for the dynamics of one's own body movements. This could explain the particular difficulty that Robert had in coordinating movements during sign production. The production of articulatory movements for signed language requires the coordination of multiple independent articulators, and the individual in this study showed a distinct deficit in combining the complex units that form the basis of sign language structure: i.e., hand configuration and targeted limb movement.

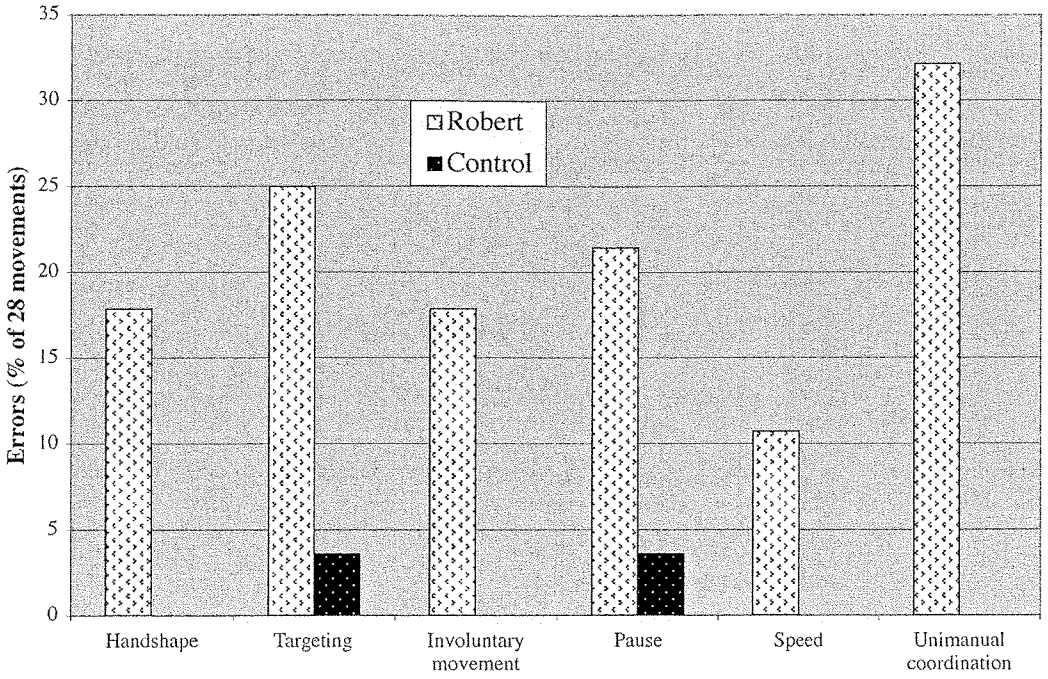

Figure 5. Pointing Task.

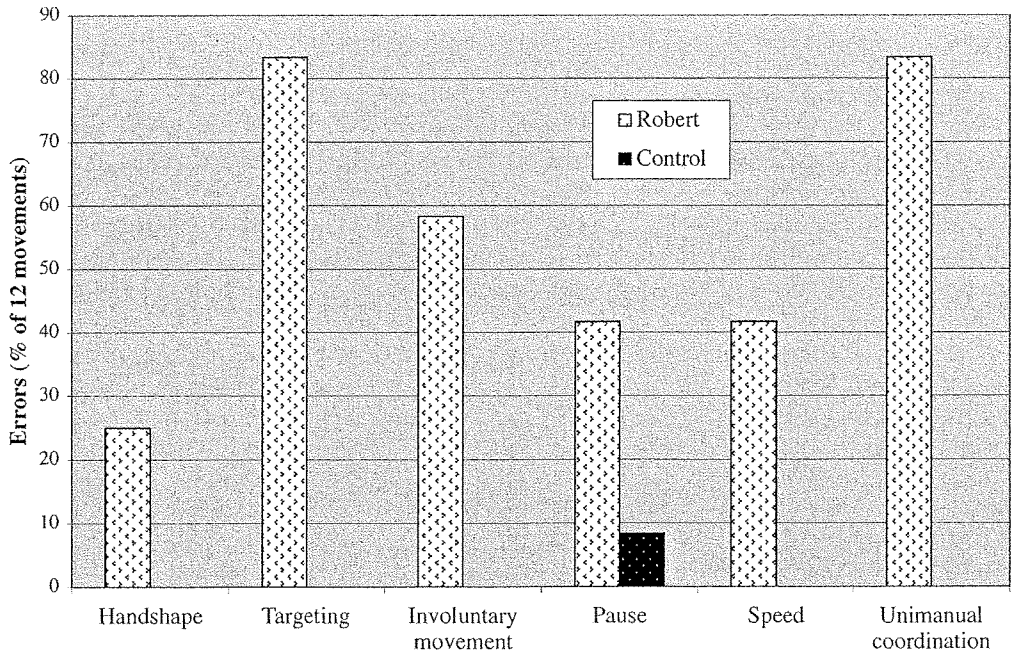

Figure 6. Reach and Grasp Task.

On the pointing task, Robert showed occasional intention tremor and consistently slow movements relative to the control. However, he showed a greater proportion of incoordination and targeting errors than involuntary movements. Furthermore, in some cases, he broke movements down into separate components and produced the components serially – a common movement anomaly in individuals with cerebellar ataxia (Zackowski et al., 2002).

On the reach and grasp task, Robert's main difficulties were with targeting and proximal/distal coordination. In addition, his hand configurations were often not well-adapted to the size and shape of the cylinders, and he produced many tremors when reaching. These findings are consistent with the results of a study by Zackowski et al. (2002), in which participants with ataxia had deficits in coordinating the components of a reach and grasp task.

Robert repeatedly used both hands on the reach and grasp task, which could be a way of compensating for tremor or task difficulty; however, this pattern is also consistent with the pattern from his signing. Whether the use of two hands for normally one-handed tasks by individuals with ataxia is common and/or voluntary is a question that should be explored in greater detail, given that experimental motor tasks are not typically designed to offer the participant the option of using one hand or two. There may be other non-linguistic tasks in which preferential bimanual movements are likely to emerge, as a result of postural instability or other motoric factors (Berrigan, Simonceau, Martin, & Teasdale, 2006). Robert had proportionally fewer targeting errors in signing than in either of the non-linguistic tasks, which parallels past findings suggesting that dysmetria occurs in non-linguistic limb movements but not in speech movements (Ackermann et al., 1997). What is novel in this study is that reaching/grasping, pointing, and language production are performed with the same articulators. Why dysmetria might occur more often in non-linguistic limb movements than in sign or speech is a question that merits further investigation. The two non-linguistic tasks examined here both have movement targets that are off the body, while the movement targets for signing are located on or in front of the body. Thus, the non-linguistic movement tasks involve allocentric movement targets, in contrast to sign or speech movements, whose targets are probably defined egocentrically.

A few qualitative observations about Robert's signing deserve mention. Robert seemed to have more difficulty producing signs which were located outside the range of his vision, for example, on his right ear or cheek. It may be that he relies heavily on vision to produce signs accurately. Further, he exhibited difficulty with signs which require a circular movement at the elbow. It is not clear whether this problem is related to dysdiadochokinesia or reflects a different type of movement deficit.

While this study is descriptive in nature, the results provide a useful comparison to previous studies of dysarthria and limb movement. The patterns that were observed on the signing and the non-linguistic tasks were consistent with the symptoms of both ataxic limb movement and ataxic dysarthria: ataxic signing was characterized by incoordination, disruptions to fine motor control, and intention tremor; and non-linguistic movements showed these characteristics in addition to dysmetria. While past research has suggested a distinction between ataxic speech and ataxic limb movements (Ackermann et al., 1997), namely with respect to dysmetria, this study did not find a consistent pattern of dysmetria in ataxic sign movements. In fact, Robert had slightly more dysmetric movements during the non-linguistic pointing task than during the signing task. This suggests that earlier findings may have been influenced as much by the task as by the effectors that were used. Signing is formationally distinct from standard motor control tasks, such as pointing or reaching, and thus may elicit a different pattern of movement deficits and be differentially affected by the motor symptoms of cerebellar ataxia. The nature of the movement task may offer some explanation of the differences in sign, speech, and non-linguistic limb movements.

This study also provides a useful comparison to past research on neurogenic disruptions to sign production (Poizner & Kegl, 1993; Tyrone, Kegl, & Poizner, 1999). In contrast to signers with movement disorders such as Parkinson's disease or apraxia, the individual in this study had large, exaggerated movements, intention tremor, and hyperextension of sign articulators. This type of movement disorder has not previously been examined in the context of sign production. Moreover, studies that have attempted to characterize the differences between linguistic and motoric sign deficits have not examined ataxia at all, but instead have focused on hypokinetic disorders.

In summary, this study illustrates that disruptions to sign articulation can take a variety of forms, in much the same way as spoken language dysarthria. Speech motor control research often examines dysarthria and disrupted limb movement as if they are unrelated phenomena with dissimilar characteristics. However, this study suggests that the form of either sign or speech dysarthria may be determined by the nature of the particular movement disorder that causes it, as well as by the set of articulators used by the language. Developing a better understanding of motor control strategies for speech and for sign will greatly advance the development of new therapies for both speech and

sign production disorders. In the field of sign language research, movement deficits have often been grouped together as a unitary phenomenon to be contrasted with linguistic deficits such as aphasia (Brentari, Poizner, & Kegl, 1995; Poizner, 1990). However, the case described here is quite different from the Deaf individuals with other movement disorders described previously, which suggests that more might be learned by examining sign language and motor deficits on a case-by-case basis. If motor deficits are treated as varied phenomena and their effects on sign language production are examined separately, we can better understand both the articulatory structure of signed languages, and the relationship between motor control, articulation and the formational properties of human language.

Original manuscript received 13 August 2008
Revised manuscript accepted 6 January 2009
First published online 6 July 2009

REFERENCES

Ackermann, H., Hertrich, I., Daum, I., Scharf, G., & Spieker, S. (1997). Kinematic analysis of articulatory movements in central motor disorders. *Movement Disorders, 12*, 1019–1027.

Atkinson, J. R., Marshall, J., Woll, B., & Thacker, A. (2005). Testing comprehension abilities in users of British sign language following CVA. *Brain and Language, 94*, 233–248.

Bastian, A. J. (2002). Cerebellar limb ataxia: Abnormal control of self-generated and external forces. *Annals of the New York Academy of Sciences, 978*, 16–27.

Battison, R., Markowicz, H., & Woodward, J. (1975). A good rule of thumb: Variable phonology in American sign language. In R. W. Fasold, & R. W. Shuy (Eds.), *Analyzing variation in language* (pp. 291–302). Washington, DC: Linstok Press.

Berrigan, F., Simonceau, M., Martin, O., & Teasdale, N. (2006). Coordination between posture and movement: Interaction between postural and accuracy constraints. *Experimental Brain Research, 170*, 255–264.

Brentari, D., Poizner, H., & Kegl, J. (1995). Aphasic and parkinsonian signing: Differences in phonological disruption. *Brain and Language, 48*, 69–105.

Brooks, V. B. (1986). *The neural basis of motor control.* New York: Oxford University Press.

Fahn, S., Greene, P. E., Ford, B., & Bressman, S. B. (1998). *Handbook of movement disorders.* Philadephia, PA: Current Medicine, Inc.

Fellows, S. J., Ernst, J., Schwarz, M., Topper, R., & Noth, J. (2001). Precision grip deficits in cerebellar disorders in man. *Clinical Neurophysiology, 112*, 1793–802.

Kent, R. D., Kent, J. F., Rosenbek, J. C., Vorperian, H. K., & Weismer, G. (1997). A speaking task analysis of the dysarthria in cerebellar disease. *Folia Phoniatrica Logopaedica, 49*, 63–82.

Kent, R. D., Kent, J. F., Duffy, J. R., Thomas, J. E., Weismer, G., & Stuntebeck, S. (2000). Ataxic dysarthria. *Journal of Speech, Language, and Hearing Research, 43*, 1275–1289.

Meier, R. P. (2002). Why different, why the same? Explaining effects and non-effects of modality upon linguistic structure in sign and speech. In R. P. Meier, K. Cormier, & D. Quinto-Pozos (Eds.), *Modality and structure in signed and spoken language* (pp. 1–26). New York: Cambridge University Press.

Poizner, H. (1990). Language and motor disorders in Deaf signers. In G. R. Hammond (Ed.), *Cerebral control of speech and limb movements* (pp. 303–326). North-Holland, NY: Elsevier.

Poizner, H., & Kegl, J. (1993). Neural disorders of the linguistic use of space and movement. In P. Tallal, A. Galaburda, R. Llinas, & C. von Euler (Eds.), *Annals of the New York Academy of Science, temporal information processing in the nervous system* (pp. 192–213). New York: New York Academy of Sciences Press.

Saigusa, H., Saigusa, M., Aino, I., Iwasaki, C., Li, L., & Niimi, S. (2005). M-mode color doppler ultrasonic imaging of vertical tongue movement during articulatory movement. *Journal of Voice, 20*, 38–45.

Stokoe, W. C. (1960). *Sign language structure: An outline of the visual communication systems of the American Deaf.* Silver Spring, MD: Linstok Press.

Timmann, D., Citron, R., Watts, S., & Hore, J. (2001). Increased variability in finger position occurs throughout overarm throws made by cerebellar and unskilled subjects. *Journal of Neurophysiology, 86*, 2690–2702.

Topka, H., Konczak, J., Schneider, K., Boose, A., & Dichgans, J. (1998). Multijoint arm movements in cerebellar ataxia: Abnormal control of movement dynamics. *Experimental Brain Research, 119*, 493–503.

Tracy, J. I., Faro, S. S., Mohammed, F. B., Pinus, A. B., Madi, S. M., & Laskas, J. W. (2001). Cerebellar mediation of the complexity of bimanual compared to unimanual movements. *Neurology, 57*, 1862–1869.

Tyrone, M. E., & Woll, B. (2008a). Sign phonetics and the motor system: Implications from Parkinson's disease. In J. Quer (Ed.), *Signs of the time: Selected papers from TISLR 2004* (pp. 43–68). Seedorf, Germany: Signum.

Tyrone, M. E., & Woll, B. (2008b). Palilalia in sign language. *Neurology, 70*, 155–156.

Tyrone, M. E., Kegl, J., & Poizner, H. (1999). Interarticulator co-ordination in Deaf signers with Parkinson's disease. *Neuropsychologia, 37*, 1271–1283.

Zackowski, K. M., Thach, W. T., & Bastian, A. J. (2002). Cerebellar subjects show impaired coupling of reach and grasp movements. *Experimental Brain Research, 146*, 511–522.