

PALILALIA IN SIGN LANGUAGE

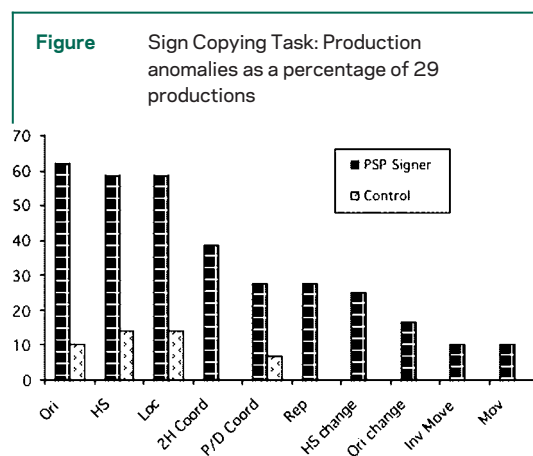


Sign languages, which use the hands and arms as articulators, are the natural languages of deaf people. Research indicates that sign, like speech, can break down at the level of motor control, in the context of a movement disorder such as Parkinson disease (PD).¹ This article describes the first known case of a deaf sign language user with progressive supranuclear palsy (PSP). PSP affects the rostral brainstem and its projections to the basal ganglia, cerebellum, and cerebral cortex.² Dysarthria typically emerges early in the course of the disease and disrupts several aspects of speech.³ The characteristic features of PSP dysarthria are articulatory incoordination and palilalia—the repetition of entire words at decreasing amplitudes without pause.² This descriptive case study suggests that those same features are prominent in the sign production of a deaf man with PSP.

Methods. Participants. The individual with PSP was a 79-year-old right-handed man, who was born deaf. He learned British Sign Language (BSL) at school at age 7, and it became his primary language. He developed left-sided weakness and dysphagia at age 77; however, no brain scan was performed. Repeated visits by researchers suggested that he had a progressive condition, and a consulting neurologist with expertise in movement disorders confirmed a diagnosis of PSP. The individual with PSP exhibited limited mobility, slow and reduced spontaneous movement, intention tremor, and stooped posture. He scored 20 out of 30 on the Mini-Mental State Examination,⁴ suggesting mild dementia; however, his BSL comprehension and production remained intact. His spoken English skills were not tested.

The man with PSP was compared on sign language measures to a healthy 70-year-old, right-handed deaf woman. The study was approved by the ethics committee at City University London. Testing procedures were explained in BSL to the participants, who gave written consent to participate. Data were collected in participants' homes by a deaf experimenter. The signing data included 29 productions of individual BSL signs, which were copied from the experimenter.

Analysis. The data were coded for four properties—handshape, location, orientation, and movement trajectory⁵—which can differentiate signs, just as consonants and vowels differentiate words. Incoordination, atypical repetitions, and involuntary movements were also coded. Deviations from the



Ori = orientation; HS = handshape; Loc = location; 2H Coord = bimanual coordination; P/D Coord = proximal/distal coordination; Rep = self repetition; HS change = handshape change; Ori change = orientation change; Inv Move = involuntary movement; Mov = trajectory of movement.

citation form of a sign were coded as anomalous productions.

Results. PSP sign productions frequently deviated from the citation form (figure). Signing anomalies occurred most often in orientation, handshape, and location. Handshape and orientation were hypoarticulated, i.e., the fingers and wrist were not fully extended, and sign locations were often lowered. In addition, the individual with PSP showed incoordination when producing two-handed signs. Other anomalies included abnormal sign repetition (palilalia) and incoordination of proximal and distal articulators on the same limb. The video (available on the *Neurology*[®] Web site at www.neurology.org) illustrates palilalic compared to normal signing.

Discussion. PSP sign movements were small, hypoarticulated, and gradual, similar to what has been reported in PD.¹ PSP signing also included involuntary movements and sign repetition, neither of which has been reported previously. The individual with PSP had incoordination across sets of articulators during signing. However, he had no difficulty with sign-internal movements that were confined to a localized set of articulators. This suggests that groups of sign articulators (e.g., the fingers) are acting as a unit, which is consistent with previous research.⁶ Sets of sign articulators probably function together as simple motor synergies, while coordination across articulators requires combination of those synergies.

Palilalia is described as the repetition of an entire word, decreasing in volume with subsequent repetitions.⁷ The disrupted signing described here is similarly palilalic, because entire signs were re-

peated, and repetitions became smaller in movement amplitude. (See the sign HOUSE, which has two internal movements and is repeated in its entirety.) However, unlike with speech, there were no observed instances of signs being repeated more than once.

Research on the forms of articulatory deficits across language modalities can lend insight into the basic articulatory units of both sign and speech. This descriptive study suggests that articulation is a modality-independent function; it consists of 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. While the case reported here is unusual, it raises larger questions for models of speech motor control. Further, it illustrates the value of reframing speech measures as broader motor control phenomena, which allows comparison to other language modalities, as well as a useful comparison to motor deficits in general.

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