available at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/cortex

Research report

Limb apraxia in corticobasal syndrome☆

Vessela Stamenova[a,*], Eric A. Roy[a,b,c,d] and Sandra E. Black[a,b,c,d,e]

[a] Department of Rehabilitation Science, University of Toronto, Canada
[b] Department of Psychology, University of Waterloo, Canada
[c] Department of Kinesiology, University of Waterloo, Canada
[d] Heart and Stroke Foundation Centre for Stroke Recovery (HSFCSR), Department of Medicine, Sunnybrook Health Sciences Centre, University of Toronto, Canada
[e] Division of Neurology, Department of Medicine, University of Toronto, Canada

ARTICLE INFO

Article history:
Received 29 July 2009
Reviewed 7 September 2009
Revised 31 March 2010
Accepted 29 April 2010
Action editor Georg Goldenberg
Published online 11 May 2010

Keywords:
Limb apraxia
Corticobasal degeneration
Corticobasal syndrome
Pantomime
Imitation
Transitive gestures
Intransitive gestures
Non-representational gestures
Skilled movement
Action
Motor control
Objects
Naming
Gesture identification

ABSTRACT

Corticobasal syndrome (CBS) is a progressive neurodegenerative disorder with asymmetric presentation and course characterized by degeneration of basal ganglia and cortical structures. Limb apraxia is a commonly observed deficit in CBS. Few studies have examined comprehensively the nature of deficits in limb apraxia. The goal of our study was to investigate the severity of deficits in various conceptual and gesture production task modalities. CBS patients were divided in two groups based on the side of brain that was initially affected by the disease. Ten patients with right hemisphere presentation (RHP) and seven with left hemisphere presentation (LHP) were included. The results showed that while selective conceptual tasks deficits were present in both groups, the overall picture suggests preserved conceptual representations of tools and actions in CBS patients with either LHP or RHP. Both groups were impaired relative to controls on gesture production tasks. Performance on transitive gestures was more severely affected in both groups than intransitive gestures. Imitation was more severely affected than pantomime, suggesting deficits in visuomotor transformations. The addition of verbal cuing during concurrent imitation affected only the LHP patients, rendering them more impaired relative to controls in their imitation with verbal cuing as opposed to their imitation only performance. Imitation of non-representational gestures was least accurate and intransitive gestures were most accurate. Patients were more severely impaired relative to controls when holding the object and when they were shown pictures of tools to pantomime.

☆ *Study carried out at:* Sunnybrook Health Sciences Centre, a University of Toronto affiliated hospital.

* *Corresponding author*. 217 Markland Cres, Nepean, ON, Canada K2G 5Z9.
E-mail address: viussi@gmail.com (V. Stamenova).

0010-9452/$ – see front matter © 2010 Elsevier Srl. All rights reserved.
doi:10.1016/j.cortex.2010.04.010

Corticobasal syndrome (CBS) is a progressive neurodegenerative disorder characterized by degeneration of basal ganglia and cortical structures. The disorder is characterized by an asymmetric presentation and course. Based on the clinical diagnostic criteria summarized by Boeve et al. (2003), CBS has an insidious onset and progressive course. The cortical deficits could present as limb apraxia, alien limb phenomenon, cortical sensory loss, neglect, constructional apraxia, myoclonus, apraxia of speech and/or non-fluent aphasia. The extrapyramidal dysfunction is characterized by focal or asymmetrical rigidity and/or dystonia. For a detailed summary of the clinical criteria and the supportive features of the clinical diagnostic criteria, the reader should consult Boeve et al. (2003).

Limb apraxia is one of the most commonly reported cortical features of CBS. It is a neurobehavioral disorder characterized by an inability to perform purposeful skilled movements, not attributable to muscle weakness, paralysis, poor comprehension, deafferentiation or an unwillingness to perform the task (Geschwind, 1975). To assess limb apraxia, patients are often asked to pantomime (e.g., perform from memory to verbal command) or imitate visually presented gestures and, thus, limb apraxia can also be operationally defined as an inability to pantomime and/or imitate gestures (Roy, 1996). This disorder is most often studied in the context of stroke and is more prevalent after left hemisphere stroke (Heilman and Rothi, 1993; Donkervoort et al., 2002; Roy et al., 1991), but limb apraxia after right hemisphere stroke has also been reported (Roy et al., 2000; Heath et al., 2001; Stamenova et al., 2010).

Several information processing models have been proposed in the literature to explain the specific patterns of deficits in patients suffering from apraxia (Geschwind, 1975; Heilman and Rothi, 1993; Goldenberg and Hagmann, 1997; Cubelli et al., 2000). The current study uses the conceptual-production systems model proposed by Roy (1996) as a framework to understand limb apraxia deficits in CBS. The conceptual-production model proposes that the execution of skilled actions is under the control of three systems: the sensory/perceptual, the conceptual and the production system. The sensory/perceptual system processes information from the environment (visual, auditory or tactile). The conceptual system stores knowledge about tools and actions. The production system consists of several subsystems subserving such functions as response selection, image generation, working memory storage of the motor plan, response organization and control of movement. Roy (1996) suggests that disruptions in any of these three systems will result in a specific pattern of praxis deficits. As an example, if the production system is affected and the conceptual system is intact, the patient should be able to recognize gestures but can neither pantomime nor imitate gestures. Thus, Roy (1996) suggested that if a patient presents with certain gesture production deficits, a comprehensive assessment examining all three systems is needed to determine where exactly disruptions in the system lie.

While limb apraxia is one of the most common cortical features of CBS, and often the initial neurobehavioral feature, few studies have examined in detail the nature of the limb praxis deficits in CBS. What we know so far, based on relatively few studies with small sample sizes, is that there appears to be greater impairment in imitation as opposed to pantomime. Peigneux et al. (2001) showed in 18 CBS patients that imitation was more impaired than pantomime, irrespective of the gesture type and two other smaller scale studies report similar findings (Jacobs et al., 1999; Spatt et al., 2002). Other series, however, have reported either no differences (Pharr et al., 2001) or the opposite effect (Leiguarda et al., 2003). In addition, studies have shown that while CBS patients improve when using actual tools relative to pantomime, they remain impaired on this task (Jacobs et al., 1999; Graham et al., 1999; Spatt et al., 2002; Leiguarda et al., 2003). Usually both transitive and intransitive gestures are affected (Leiguarda et al., 2003; Jacobs et al., 1999; Peigneux et al., 2001; Buxbaum et al., 2007), but some report greater impairments on transitive than intransitive gestures (Pharr et al., 2001; Salter et al., 2004; Chainay and Humphreys, 2003). Further, no differences between representational and non-representational gestures have been found (Merians et al., 1999; Spatt et al., 2002; Salter et al., 2004; Leiguarda et al., 2003; Buxbaum et al., 2007). Finally, the little evidence that exists to date with respect to conceptual tasks suggests that most patients with CBS do not have impairment in their conceptual knowledge of actions and tools (Leiguarda et al., 1994; Jacobs et al., 1999; Graham et al., 1999; Soliveri et al., 2005).

Most studies to date examining apraxia in CBS suffer from several methodological downfalls. First, most reports have included very few patients. In fact, in a recent review, we reported that out of the 16 studies examining apraxia deficits in the literature, only six included more than five CBS patients (Stamenova et al., 2009). In addition, most investigations assess patients only on a few task modalities. Finally, Roy (1996) differentiates between delayed and concurrent imitation. In concurrent imitation the examiner demonstrates an action until the patient completes the imitation, while in delayed imitation the examiner demonstrates an action and the patient imitates the examiner right after from memory. According to Roy (1996), comparing the performance in these two conditions helps determine whether deficits in the production system stem from deficits in analysis of visual gestural information (both concurrent and delayed imitation impaired), in encoding visual gestural information into working memory (selective impairment in delayed imitation) or in deficits of response organization and control (impairments in both imitation conditions as well as in pantomime). No studies to date have compared CBS patients' performance on these two tasks.

Thus, the goal of our study was to examine comprehensively limb apraxia deficits in a relatively large sample of 17 CBS patients. Transitive, intransitive and non-representational gestures were examined. Patients were assessed in pantomime, object use, delayed and concurrent imitation, as well as conceptual knowledge. Based on past literature, the following predictions are made. First, if differences between pantomime and imitation were to be observed, performance on imitation should be more impaired than pantomime. Pantomiming when holding the tool (object/tool use) would cause less severe impairment than pantomiming. Performance on transitive gestures would be more impaired than intransitive gestures. Even though past studies have not