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Communication and pragmatic breakdowns in amyotrophic lateral sclerosis patients

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ABSTRACT

While there is increasing attention toward cognitive changes in amyotrophic lateral sclerosis (ALS), the domain of pragmatics, defined as the ability to integrate language and context to engage in successful communication, remains unexplored. Here we tested pragmatic abilities in 33 non-demented ALS patients and 33 healthy controls matched for age and education through 6 different tasks, ranging from discourse organization to the comprehension of figurative language, further grouped in three composite measures for pragmatic production, pragmatic comprehension and global pragmatic abilities. For a subgroup of patients, assessment included executive functions and social cognition skills. ALS patients were impaired on all pragmatic tasks relative to controls, with 45% of the patients performing below cut-off in at least one pragmatic task, and 36% impaired on the global pragmatic score. Pragmatic breakdowns were more common than executive deficit as defined by the consensus criteria, and approximately as prevalent as deficits in social cognition. Multiple regression analyses support the idea of an interplay of executive and social cognition abilities in determining the pragmatic performance, although all these domains show some degree of independence. These findings shed light on pragmatic impairment as a relevant dimension of ALS, which deserves further consideration in defining the cognitive profile of the disease, given its vital role for communication and social interaction in daily life.

1. Introduction

Research in the last decades has radically modified the traditional view of amyotrophic lateral sclerosis (ALS) as a pure motor neuron disease, leading to the recognition of cognitive and behavioral changes as an integral feature of the disease, in addition to motor manifestations (Goldstein & Abrahams, 2013; Phukan et al., 2012). Several studies indicated that cognitive impairments affect approximately 50% of the ALS population, with a spectrum ranging from pure motor disorders to dementia, most often of the fronto-temporal type (Bennion Callister & Pickering-Brown, 2014; Consonni et al., 2013; Montuschi et al., 2014). In this continuum, a large proportion of non-demented ALS patients (approximately 30%) have cognitive dysfunctions, predominantly dysexecutive syndrome, with the earliest and most commonly reported signs consisting in deficit in verbal fluency, followed by other features, such as disturbances in problem solving, attentional control and reasoning.

In addition to the executive deficit, impairments in social cognition are frequently reported in ALS (Abrahams, 2011). Patients perform poorly in processing emotions and in attributing emotional and cognitive states to others, as assessed through a variety of tasks of different complexity, from eye-gaze to cartoon stories and social decision making (Cecchetto et al., 2014; Cerami et al., 2014; Palmieri et al., 2010; van der Hulst, Bak, & Abrahams, 2014). Notably, impairments in social cognition seem to be more diffuse and possibly dissociated from executive dysfunction (Girardi, Macpherson, & Abrahams, 2011).

Other studies have investigated linguistic aspects, both in production and comprehension. Independently of speech articulation problems, patients might have impaired sentence grammar (Ash, Olm et al., 2014) and single word processing (Leslie et al., 2014), especially for verbs as compared to nouns (Bak & Hodges,

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2004; Papeo et al., 2015). Impaired performance in an extensive battery of tests assessing different aspects of language processing has been reported in about 40% of the non-demented ALS population (Taylor et al., 2013). Executive deficit does not fully account for these linguistic breakdowns, similarly to what observed for social cognition (Abrahams, 2013).

While deficits in executive functions, social cognition and selected aspects of language are well documented, one domain that is nearly unexplored in ALS is pragmatics, i.e. the ability of integrating language and context for the purpose of the communicative exchange (Bambini, 2010; Bambini & Bara, 2012). When used in naturalistic environments and social interaction, language is more than coding and decoding words and sentences, and involves the recognition of the speaker's communicative intention based on contextual clues, as well as engaging in contextually appropriate discourse and conversation (Grice, 1975; Sperber & Wilson, 1995). The complexity of these processes becomes especially evident in language uses that massively exploit context, such as non-literal language. A large amount of research in neuropragmatics has documented deficits in dealing with contextual aspects of language, first described in right-hemisphere brain damaged patients, and later observed in many clinical populations, including different forms of dementia, schizophrenia and autism (Bambini & Bara, 2012; Stemmer, 2008). Despite relatively intact abilities in formal aspects of language (i.e. word and sentence processing), these patients typically have difficulties in understanding metaphors and idioms, in deriving ironic and humoristic nuances, in producing discourse that stays on topic and is appropriate to the conversational context, all which makes communication with familiars and proxies especially difficult.

There are several reasons to hypothesize a pragmatic impairment in ALS. First, failures in organizing discourse (Ash et al., 2006; Roberts-South, Findlater, Strong, & Orange, 2012) and in comprehending non-literal expressions such as metaphors (Orange & Hillis, 2012) have been reported in fronto-temporal dementia (FTD). Given the notion of an FTD–ALS spectrum (Bennion Callister & Pickering-Brown, 2014), it is not surprising that discourse analysis has revealed similar modifications in the speech produced by ALS patients (Ash, Menaged et al., 2014), and impairment is likely to extend to other pragmatic domains.

Second, although to a degree that varies between different clinical populations, pragmatics seems to be connected to other cognitive abilities, especially social cognition and executive functions (Martin & McDonald, 2003), that are known to be impaired in ALS. For instance, the association between pragmatics and social cognition is especially evident in tasks that involve the interpretation of linguistic materials based primarily on intention attribution, such as sarcasm detection. In similar tasks, patients with behavioral variant of FTD perform poorly (Kipps, Nestor, Acosta-Cabronero, Arnold, & Hodges, 2009; Shany-Ur et al., 2011), and so do ALS patients (Staios et al., 2013). Impairments in ALS might extend also to other pragmatic tasks that capitalize on social cognition abilities. As for executive functions, they form a system that coordinates behavior and enables individuals to use their cognitive abilities in a flexible manner through different situations, which represents an important platform for adapting to the communicative context of the ongoing conversation (Martin & McDonald, 2003). The dysexecutive syndrome observed in ALS might thus be associated to pragmatic impairment as well.

Third, anatomical basis of pragmatics are also compatible with descriptions of structural and functional abnormalities in ALS. Pragmatic abilities are supported by frontal and temporal networks in both hemispheres. In addition, pragmatics engages mentalizing regions such as the medial prefrontal cortex and the temporo-parietal junction (Catani & Bambini, 2014; Hagoort & Levinson, 2014). Functional neuroimaging in ALS has frequently described frontotemporal dysfunction in relation to dysexecutive deficit (Tsermentseli, Leigh, & Goldstein, 2012), as well as malfunctioning in dorsolateral and medial prefrontal cortex (Girardi et al., 2011), including alteration of functional asymmetry (Palmieri et al., 2010), possibly related to emotional and social cognition deficits. Other studies have reported reduction of white matter integrity in the frontal, temporal and parietal lobes, including frontotemporal-limbic connections important for social cognition (Crespi et al., 2014). Given this pattern of impairment, the circuits underlying pragmatics may be affected as well.

The primary aim of this study was to explore for the first time the domain of pragmatic abilities in non-demented ALS patients, with the ultimate goal of contributing to the description of the cognitive profile of the disease in order to include this important domain of language and social interaction. Secondly, we aimed at investigating the relationship between pragmatic behavior and performance on executive functions and social cognition tasks. Based on previous evidence on pragmatic impairment in FTD and given the notion of ALS–FTD continuum, we hypothesized that pragmatic deficit might affect a large percentage of non-demented ALS population. Finally, we hypothesized that this impairment might be intertwined with social cognition and dysexecutive deficits.

2. Materials and methods

2.1. Participants

The patient group consisted of thirty-three non demented sporadic ALS patients (13 females; mean age, years ± standard deviation: 63.30 ± 9.64; mean education: 9.79 ± 4.61) recruited through the General Neurology Department of National Neurological Institute 'Casimiro Mondino', Pavia. All of them were native speakers of Italian and had a diagnosis of probable or definite ALS based on Revised El Escorial Criteria (Brooks, Miller, Swash, & Munsat, 2000) and electrodiagnostic criteria (de Carvalho et al., 2008). None of the patients met consensus criteria for diagnosis of frontotemporal dementia (Neary et al., 1998). Other exclusion criteria were major comorbid medical, neurological or psychiatric history. Patients scored 34.85 ± 8.83 (range 0–48) on the ALS Functional Rating Scale (Cedarbaum et al., 1999). Disease onset frequencies were as follows: bulbar ($n = 8$; 24.24%), limb ($n = 25$; 75.76%), consistent with typical phenotype frequencies. Mean disease duration, defined as $\text{date}_{\text{time examination}} - \text{date}_{\text{start ALS symptoms}}$ was 25.61 ± 18.99 months. See Table 1 for the population details.

The control group consisted of thirty-three neurologically healthy adults (18 females; mean age: 61.24 ± 6.66; mean education: 10.97 ± 4.34), all native speakers of Italian. The ALS patients and the controls did not significantly differ in terms of age ($p = 0.32$) or years of education ($p = 0.29$).

The study was approved by the local Ethics Committee. Informed consent was obtained from all participants in accordance with the principles of the Declaration of Helsinki.

2.2. Pragmatic assessment

Pragmatic abilities were assessed in ALS patients ($n = 33$) and in controls ($n = 33$) through a novel test (APACS, Assessment of Pragmatic Abilities and Cognitive Substrates) combining different pragmatic tasks that are widely used in the literature, with refined linguistic materials in Italian, and in a unified framework inspired by language pragmatics (Grice, 1989; Sperber & Wilson, 1995). Two pragmatic domains were targeted, namely discourse and non-literal meanings, as they are widely impaired in clinical conditions. Special care was taken to preserve the contextual validity of the linguistic materials, and to control for psycholinguistic