Language reconfiguration in bilinguals: a study with Huntington's disease patients

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Abstract

The present study investigated language inhibition and cross-language interference as two possible mechanisms of bilingual language control (BLC) that can be affected by Huntington's disease (HD), a neurodegenerative disease (ND) affecting the striatum. To this aim, the study explored the performance of pre-symptomatic and early-stage HD patients in two experimental tasks meant to elicit cross-language interference and language inhibition, including a Stroop task and a language switching task. The results revealed dissociations between these two mechanisms, indicating that language activation or inhibition is related to HD pathology while cross-language interference is not. Switch costs in HD patients were greater than controls in low-demand control conditions of language switching (longer preparation time), while Stroop effects were similar between the two groups of participants. This result was interpreted as a difficulty in overcoming the excessive inhibition applied to non-target language. The BLC processes related to the striatum and subcortical structures are discussed.

Keywords: bilingualism, movement disorders, language control, inhibition, cross-language interference

1. Introduction

Interest in research on language and cognition in bilingual speakers with neurodegenerative diseases (NDs) is increasing. This interest is motivated by the high prevalence of age-related disorders as a consequence of longer life expectancy. At the same time, bilingualism and multilingualism are also on the rise, and this situation raises many questions regarding the interaction between NDs and bilingualism. Research on bilingual language processing in patients with NDs offers an excellent opportunity to complement data on brain damage with data on healthy individuals (Calabria, Cattaneo, & Costa, 2017).

One issue explored in this context is the study of the architecture of language control in bilingual people. Bilingual language control (BLC) is defined as the skill that enables bilinguals to use a target language while monitoring for potential interference from the language not in use (Green, 1986, 1998). In the context of NDs, BLC processes have been explored in brain diseases affecting the executive control (EC) system, such as Parkinson's disease (PD) (Cattaneo et al., 2015). The results of this research revealed that basal ganglia degeneration may affect language control, even before the EC system is significantly impaired, at least for some processes related to inhibitory control. These results suggest that research on bilingual patients with basal ganglia degeneration offers an opportunity to gain a greater understanding of BLC processes related to these brain structures.

The present study explores how the degeneration of the striatum, as a consequence of Huntington's disease (HD), affects two processes involving BLC: cross-language interference and language activation/inhibition. These two processes have been related to striatum structures (caudate nuclei and putamen) in healthy bilinguals who are required to switch between languages (Abutalebi et al., 2008; 2013), and these processes are included in the BLC network as crucial structures for language selection (Abutalebi & Green, 2008, 2016; Calabria, Costa, Green, & Abutalebi, 2018; Luk, Green, Abutalebi, & Grady, 2011). The study assesses whether these BLC processes are affected in HD patients as a result of the degeneration of the striatum.

Executive control impairments are present in HD patients from the early onset of disease and even in pre-symptomatic stages. Some studies have reported EC deficits in switching abilities

(Aron et al., 2003), attention (Maurage et al., 2017; Thompson et al., 2010), conflict monitoring and inhibitory control (Baake, Reijntjes, Dumas, Thompson, & Roos, 2017; Lemiere, Decruyenaere, Evers-Kiebooms, Vandenbussche, & Dom, 2004; Paulsen et al., 2014; Peinemann et al., 2005). Recent studies have found a clear correlation between EC deficits in HD and the hypometabolism of the striatum, one of the primary subcortical structures affected in HD patients (e.g., Georgiou-Karistianis et al., 2013; Montoya, Price, Menear, & Lepage, 2006; Paulsen et al., 2014; Rosas et al., 2006; Vandenberghe, Demaerel, Dom, & Maes, 2009; Vandervert, 2016). Also, the degree of degeneration of this brain area predicts the longitudinal clinical outcomes in HD patients and the cognitive decline over time (Domínguez et al., 2017; Tabrizi et al., 2013). Essentially, the pathology affecting the striatum leads to an excessive inhibition which is counterproductive when performing EC control tasks (Aron et al., 2003) and response selection in particular (Aron et al., 2003; Lawrence, Sahakian, & Robbins, 1998; Smith & Shadmehr, 2000).

While EC deficits have consistently been found in HD patients, currently, no evidence links these deficits to language control. Huntington's disease patients have deficits in language learning (De Diego-Balaguer et al., 2008), verb naming (Kargieman et al., 2014), articulatory planning for speech (Skodda et al., 2016), and syntactic and sentence processing (Hinzen et al., 2017; Teichmann, Gaura, et al., 2008; Teichmann, Darcy, Bachoud-Lévi, & Dupoux, 2009; Teichmann, Dupoux, Cesaro, & Bachoud-Lévi, 2008; Ullman et al., 1997). Given that pathologies with degeneration in the same subcortical network, such as PD, induce deficits in BLC processes (Cattaneo et al., 2015), this relationship may suggest that basal ganglia play a role in language control as well.

1.1. The role of subcortical structures in the BLC network

The striatum, and specifically the caudate nuclei, is described as an essential part of the BLC network that also includes the dorsolateral prefrontal cortex, supplementary motor area, anterior cingulate cortex and cerebellum (Abutalebi & Green, 2007, 2016; Hervais-Adelman, Moser-Mercer, & Golestani, 2011; Luk et al., 2011; Pliatsikas & Luk, 2016; Seo, Stocco, & Prat, 2018). According to Abutalebi and Green (2007), the striatum is involved in selecting and

controlling the activation or inhibition of the language in use. Moreover, some studies have highlighted that the activation of the prefrontal cortex and striatum is modulated by the linguistic context in which bilinguals are communicating. For instance, Abutalebi et al. (2008) have found that the left caudate was activated when participants were required to switch languages (dual-language context), but not when participants had to switch intra-language (naming pictures as objects or actions in a single-language context). Similarly, the activation of the left caudate occurs only in cross-language priming conditions. Crinion et al. (2006) presented sequential word pairs and instructed bilinguals to judge the pairs' semantic relatedness. As a key part of the experiment, the target word was either in the same or a different language. The results revealed an increased activation of the left head of the caudate when word pairs were in different languages (dual-language context) but not when they were in the same language (but see also Ali, Green, Kherif, Devlin, & Price, 2010, for a more general effect of verbal interference and caudate). Moreover, the crucial role of this area in language control has been demonstrated in lesion and brain stimulation studies. That is, electrical stimulation of (Wang, Wang, Jiang, Wang, & Wu, 2012) and brain damage to the left caudate generate pathological language switching and mixing (Abutalebi, Miozzo, & Cappa, 2000; Aglioti, Beltramello, Girardi, & Fabbro, 1996; Aglioti & Fabbro, 1993; Ansaldi, Saidi, & Ruiz, 2010; Calabria, Marne, Romero-Pinel, Juncadella, & Costa, 2014; Fabbro, Skrap, & Aglioti, 2000; Garcia-Caballero et al., 2007; Kong, Abutalebi, Lam, & Weekes, 2014; Leemann, Laganaro, Schwitser, & Schnider, 2007; Mariën, Abutalebi, Engelborghs, & De Deyn, 2005).

Caudate nuclei (and frontostriatal circuits) have a direct role in the inhibition of inappropriate behaviours (Aron, 2011; Casey, Durston, & Fossella, 2001; Rao et al., 2014) and of non-target language in bilingual production (Abutalebi & Green, 2016; Abutalebi & Green, 2007). Language activation and inhibition are control abilities that are essential to a bilingual's ability to switch between languages. At the behavioural level, language inhibition is usually measured by eliciting language switching costs, which are the increase in naming latencies that occurs when bilinguals name a picture after switching languages. Evidence suggests that the language switch cost is greater in bilingual patients with basal ganglia degeneration (as in PD)

compared to healthy controls, but not when the same patients perform non-linguistic task switching (Cattaneo et al., 2015). These results suggest that the basal ganglia play a special role in BLC. Abutalebi and Green (2016) have proposed that the left caudate nucleus is involved in dual-language contexts and its activation likely reflects the burden associated with language selection processes. In their revised model of BLC, Abutalebi and Green (2016) described the caudate nuclei as an access gate that regulates the activations of both the prefrontal cortex and posterior cortical regions in dual-language contexts.

Given that caudate nuclei are one of the main subcortical structures affected in HD, deficits in BLC may be expected as well. Regarding EC deficits in HD patients, it has been proposed that striatum degeneration would lead to excessive inhibition control (Aron et al., 2003) or impaired response selection (Aron et al., 2003; Lawrence, Sahakian, & Robbins, 1998; Smith & Shadmehr, 2000). In observing BLC, deficits in language inhibition are expected in HD patients, as proposed by neural models of BLC (Abutalebi & Green, 2008, 2016).

1.2. The present study

We explore HD patients' performance in two tasks that reveal cross-language interference and language activation/inhibition: the Stroop task and the language switching task.

We use Stroop task to assess the integrity of the lexical retrieval system and the efficiency in preventing cross-language interference in HD patients. Participants are asked to name the text colour of words written in their dominant-language while ignoring the words' meaning. In the within-language condition, the words are presented in the same language in which the response has to be given (dominant language); in the between-language condition, the words are presented in the non-dominant language of the participants. The performance of HD patients in this task is a strong predictor of the progression from the pre-manifestation to the manifest stage of the disease (Baake, Reijntjes, Dumas, Thompson, & Roos, 2017). Therefore, it is expected that HD patients would perform worse than healthy controls in both conditions. Additionally, it is predicted that patients would display a larger Stroop effect (semantic interference) in the between-

rather than within-language condition as compared to controls. This prediction is based on evidence that the left caudate is more sensitive to conditions in which bilinguals must manage two languages (e.g., Abutalebi et al., 2008; Crinion et al., 2006). Hence, considering that HD patients suffer from damage to this structure, one would expect the between-language condition to be especially affected.

We use language switching to investigate the integrity of inhibitory control related to language reconfiguration/reactivation. Specifically, we compare the individual performance on two versions of the language switching task which differ in one parameter, the cue-stimulus interval (CSI). This experimental manipulation is introduced to see whether the reconfiguration of a new language is influenced by the time needed for inhibition to dissipate. In one version of the task, the cue for the naming language is presented simultaneously with the picture to be named (short CSI) whereas, in a second version, the cue is presented one second before the actual picture (long CSI). In comparing these two task designs, previous studies have demonstrated that the magnitude of the switch costs tends to be larger with short CSIs and is reduced as CSIs increase (Costa & Santesteban, 2004; Fink & Goldrick, 2015; Ma, Li, & Guo, 2016; Mosca & Clahsen, 2016; Verhoef, Roelofs, & Chwilla, 2009). This reduction is thought to index people's ability to overcome inhibition of the non-target language in changing the current language. That is, if people are given more time between the cue signalling a new naming language and the picture to name (long CSI), they experience less of a switch cost. This suggests that the inhibition applied to the previous language has already disappeared and it no longer is able to interfere with production of the new language.

In undertaking this experiment, our hypothesis is that HD patients will need longer period of time to override the effect of inhibition from the previous language. This is based on the evidence that HD patients would have less efficient inhibitory control, as shown in larger switch costs with long CSIs while performing non-linguistic task switching (Aron, Watkins, et al., 2003). Similarly, we expect that HD patients will not benefit as much as healthy controls from this longer preparation time and that, consequentially, they will show similar switch costs for both CSIs.

In the language switching task, we also explore the so-called mixing costs, an additional measure of language control abilities (see Cattaneo et al., 2015 for evidence with bilingual PD patients). This cost is calculated by subtracting the naming latencies of the trials presented in a block-design language naming condition from those of the repeat trials (in the mixed condition). This measure is introduced to report a more general and sustained control of BLC (“proactive control”, Cattaneo et al., 2015). We expect that, if this type of control is not specifically related to language activation/inhibition, the magnitude of mixing costs would be the same in HD patients and healthy controls. Furthermore, HD patients would show a similar decrease in mixing costs as a function of preparation time when compared with healthy controls (Ma et al., 2015). Therefore, we predict that proactive control would not be affected in HD patients as not directly related to basal ganglia activity (e.g., De Pisapia & Braver, 2006).

In the current study, we do not include neuroimaging data as our hypotheses are based on findings from other studies that the subcortical structures (specifically the striatum) are affected from the onset of the pre-symptomatic stage of HD (Georgiou-Karistianis et al., 2013; Rosas et al., 2006; Vandenberghe et al., 2009). Therefore, damage to the striatum in our HD sample is inferred based on this evidence along with the presence of symptoms (e.g., motor impairments) that are clinical markers of the dysfunction of this subcortical area.

2. Methods

2.1. Participants

Twelve bilingual individuals with positive genetic testing (CAG repeats mean = 42.75 ± 1.5 , 41-46) for HD (10 females, mean age = 42.2 ± 5.5 , mean education = 13.9 ± 1.9) and fourteen healthy controls matched for age and education (10 females, mean age = 41.9 ± 4.1 , $p = 0.87$; mean education = 14.6 ± 1.7 , $p = 0.31$) participated in this study. All individuals with HD were recruited at the Movement Disorders Unit of the Hospital de la Santa Creu i Sant Pau in Barcelona, diagnosed by an experienced neurologist and evaluated using the Unified Huntington's Disease Rating Scale (UHDRS; Kiebertz, 1996). Eight of 12 participants were in the pre-symptomatic stage of the disease (UHDRS score ≤ 4 , Diagnostic confidence level < 4). Symptomatic patients

had a mild degree of motor impairments according to the UHDRS (13.17 ± 18.2). On the total functional capacity (TFC) scale, HD patients were largely functional ($12.33 \pm .8$, 11-13) with all falling within early-stage or stage I HD according to Shoulson et al. (1989). Neuropsychiatric symptoms were also present to a minor extent in the HD group (apathy= 3.8 ± 4.8 ; depression= 6.0 ± 7.1 ; irritability= 4.5 ± 6.4) and no patient had any psychotic disorders. As part of the UHDRS, the Symbol Digit Modality Test, Stroop Colour Word Test and a verbal fluency test were included to measure cognitive deficits. The scores on cognitive tests were comparable to those of pre-symptomatic or early-stage HD patients in previous studies (e.g., De Diego-Balaguer et al., 2008; Hinzen et al., 2017) (Symbol Digit Modality Test = 45.30 ± 15.8 , Stroop Colour Word Test = 42.2 ± 12.77 , verbal fluency = 22.40 ± 4.32).

As for the clinical symptoms of the disease, symptomatic patients were significantly different from pre-symptomatic in their motor score on the UHDRS ($p < .01$), but not in the cognitive score ($p = .09$). Additionally, the degree of neuropsychiatric symptoms ($p_s > .25$) and disease burden ($p = .11$) were not found to be statistically different between the two groups.

2.1.1. Linguistic profile

Language history and dominance were determined by means of a questionnaire administered to participants as well as interviews conducted during the experimental session. Each participant self-rated their proficiency in two languages – Catalan and Spanish – on a four-point scale of their speaking, comprehension, writing and reading abilities (1 = poor, 2 = regular, 3 = good, 4 = perfect) (See Appendix). As illustrated in Table 1, both patients and healthy controls were highly proficient in all four linguistic domains. Participants were considered to be early bilinguals as, on average, they had been regularly exposed to their non-dominant language starting at 6 years old. Age of acquisition was not significantly different between the two languages ($p > .05$).

Language use was examined by means of ten questions in which participants were required to report how frequently they spoke the two languages during their lifetime; scores were transformed into percentages, with 0% being equivalent to using only Spanish, 100% being

equivalent to using only Catalan, and 50% being equivalent to balanced use of the two languages. Both patients and healthy controls reported using Catalan and Spanish in with equal frequency (57.3% and 56.3% respectively). Given that all participants were early bilinguals and reported similar proficiency in both languages, no objective criteria were used to define individual language dominance. The preference expressed by participants when asked which language they felt more comfortable speaking was used as a proxy. Seven HD patients and seven healthy controls reported greater dominance in Spanish; the remaining participants reported greater dominance in Catalan.

INSERT TABLE 1 ABOUT HERE

2.2. Materials and procedure

All participants were tested in the two versions of language switching, one with short and one with long CSI, as well as in within-language and between-languages versions of a Stroop task. In most cases, the tasks were administered in one experimental session. When time constraints posed a challenge, the tasks were run during two sessions a week apart (for 9 out of 12 HD patients). All tasks were administered using a laptop (screen 15.6" and resolution of 1280 x 800) and controlled with the DMDX software (Forster & Forster, 2003). Responses were analysed offline, and naming latencies were measured using the Checkvocal software (Protopapas, 2007).

2.2.1. Language switching

Two sets of eight pictures of objects (with non-cognate names) were selected from Snodgrass and Vanderwart (1980). The two sets were designed such that different pictures appeared within the two versions of the language switching task (short vs. long CSI).

In each task, participants were required to name eight pictures in Catalan or in Spanish as quickly as possible according to a cue (the Catalan or the Spanish flags). The pictures were presented in single and mixed blocks. In the single blocks, the naming language (Spanish or

Catalan) was held constant for all trials; in the mixed blocks, participants had to name the pictures according to the flag cue on the screen. The mixed blocks include two types of trials: repeat trials in which participants had to name objects in the same language used in the previous trial; and switch trials in which participants were required to name objects in the language other than the one used in the previous trial (see Cattaneo et al., 2015).

Each of the two versions of the task included a total of 96 trials in the single block condition and 96 trials in the mixed block condition (33 Spanish repeat trials, 33 Catalan repeat trials, 15 Spanish switch trials and 15 Catalan switch trials). The proportions of switch and repeat trials were 31% and 69%, respectively. Every trial began with a fixation point (a white cross) in the centre of the screen displayed for 500 ms, followed by a cue (a Spanish or Catalan flag) and then by the picture for a maximum of 2500 ms. In the short CSI version of the task, the cue and target appeared at the same time (CSI 0); in the long CSI version, the cue was displayed for 1000 ms (CSI 1000), and then the picture appeared for a maximum of 2500 ms.

2.2.2. Stroop task

Nine adjectives in Spanish and Catalan were selected as distractor words, and three ink colours were included in each language (green, red and yellow). Participants were required to name the colour in which the words were printed, and each colour was presented with three types of distractor words. In the within-language condition, the ink colour was named in the participant's dominant language (Spanish or Catalan depending on each participant), and the distractor word belonged also to the dominant language (see Participants section for the classification of language dominance). In the between-language version, the distractor words were in the non-dominant language and the ink colour was named in the participant's dominant language.

The three conditions for distractors were as follows: 1) non-colour words printed in yellow, red or blue, called the "neutral" condition; 2) names of the colours printed with a congruent ink colour, the "congruent" condition; and 3) names of the colours printed with a

different ink colour, the “incongruent” condition. Green ink was used as a filler and was presented in combination with the words of the three conditions.

Each trial was organized as follows: a fixation point was presented for 1000 ms, then a word appeared on the screen for a maximum of 2000 ms. For each version of the task, each word-colour combination was presented four times for a total of 96 trials (72 experimental and 24 fillers). Each experimental session began with a block of ten practice trials.

3. Statistical analysis

Both naming latencies and accuracy were analysed. Latencies exceeding three standard deviations above or below a participant’s mean and incorrect responses were excluded from the analyses. Verbal responses were classified as errors in the following cases: “omission”, when a participant was unable to name an object; “lexical”, when a participant produced a word that was not semantically or phonologically related to the target; “cross-language intrusion”, when a participant produced the correct word but in the non-requested language; and “phonological”, when the participant deleted, substituted or added phonemes to the correct word related to the picture.

The language switching data was analysed with a repeated measures ANOVA with type of trial (single, repeat, and switch), language (dominant, non-dominant), and CSI (short, long) as within-subject factors, and group (HD patients, healthy controls) as between-subject factors.

The Stroop task data was analysed with a repeated measures ANOVA with type of trial (neutral, congruent and incongruent) and language context (within-language, between-languages) as within-subject factors, and group (HD patients, healthy controls) as between-subject factors.

Both F_1 (by-participants) and F_2 (by-items) values are reported within the statistics performed for the language switching task and the Stroop task.

4. Results

4.1. Language switching

Naming latencies. The omnibus ANOVA revealed differences between the two versions of the language switching task as illustrated by a significant main effect of CSI [$F_1(1, 24) = 16.16$, $p < .001$, $\eta^2 = .40$; $F_2(2, 14) = 16.77$, $p < .01$, $\eta^2 = .54$] and interaction between CSI and type of trial [$F_1(2, 48) = 8.11$, $p < .01$, $\eta^2 = .25$; $F_2(2, 28) = 3.34$, $p < .05$, $\eta^2 = .19$]. Moreover, the significant interaction between type of trial and group [$F_1(2, 48) = 6.35$, $p < .01$, $\eta^2 = .21$; $F_2(2, 28) = 5.89$, $p < .05$, $\eta^2 = .29$] suggested different patterns (or magnitude) of switching costs between patients and healthy controls. To explore these different patterns, each task version was analysed separately.

4.1.2. Short CSI (0 ms)

Naming latencies. The results revealed a significant main effect of type of trial [$F_1(2, 48) = 61.59$, $p < .001$, $\eta^2 = .72$; $F_2(2, 28) = 47.79$, $p < .001$, $\eta^2 = .77$]. Naming latencies for three conditions were significantly different among them (single trials: 912 ms; repeat trials: 1000 ms; switch trials: 1078 ms; all $p_s < .05$) indicating the presence of both switch and mixing costs (see Figure 1). The main effect of group was also significant [$F_1(1, 24) = 9.01$, $p < .01$, $\eta^2 = .27$; $F_2(1, 14) = 21.44$, $p < .001$, $\eta^2 = .60$] indicating that HD patients were slower (1121 ms) than healthy controls (872 ms). No other interactions or main effects were statistically significant.

Accuracy. The results revealed a significant main effect of type of trial [$F_1(2, 48) = 6.28$, $p < .01$, $\eta^2 = .21$; $F_2(2, 28) = 21.44$, $p < .001$, $\eta^2 = .60$], and post-hoc analyses revealed that accuracy was lower for switch (95.9%) and repeat (96.5%) trials compared to single trials (99.0%, $p < .05$). The main effect of language was significant only in the by-participants analysis [$F_1(1, 24) = 4.49$, $p < .05$, $\eta^2 = .16$; $F_2(2, 28) = .84$, $p = .38$] indicating that accuracy was slightly higher in the dominant (97.8%) than the non-dominant language (96.5%). Finally, HD patients (94.5%) were less accurate than healthy controls (99.8%) [$F_1(1, 24) = 5.56$, $p < .05$, $\eta^2 = .18$; $F_2(1, 14) = 5.42$, $p < .05$, $\eta^2 = .28$], and their performance was modulated by the type of trial [group x type of trial interaction: $F(2, 48) = 5.72$, $p < .01$, $\eta^2 = .19$; $F_2(2, 28) = 3.25$, $p = .05$, $\eta^2 = .19$]. Post-hoc

analyses revealed that HD patients were less accurate in switch (92.2%) than single trials (98.1%, $p < .05$) while healthy controls performed with the same level of accuracy in all type of trials.

4.1.3. Long CSI (1000 ms)

Naming Latencies. The results revealed a significant main effect of type of trial [F_1 (1, 48) = 16.88, $p < .001$, $\eta^2 = .41$; F_2 (2, 28) = 21.72, $p < .001$, $\eta^2 = .61$], and post-hoc analyses showed that naming latencies were fastest for single trials (877 ms), slowest for switch trials (959 ms) and trials in the middle for repeat (916 ms) ($p_s < .05$). Moreover, the main effect group [F_1 (1, 24) = 10.90, $p < .01$, $\eta^2 = .31$; F_2 (1, 14) = 4.53, $p = .05$, $\eta^2 = .21$] and the interaction between group and type of trial [F_1 (1, 48) = 3.58, $p < .05$, $\eta^2 = .13$; F_2 (2, 28) = 4.05, $p < .05$, $\eta^2 = .22$] were significant. No other main effects or interactions were statistically significant.

The significant interaction between group and type of trial suggests differences in the magnitude of switch and mixing costs between patients and healthy controls. To further assess these potential differences, we used proportional costs to account for group differences in speed of processing. Proportional switch costs were calculated as the difference between naming latencies in switch trials and repeat trials (mixed blocks) divided by naming latencies in repeat trials. Likewise, proportional mixing costs were calculated as the difference between naming latencies in repeat and single trials divided by naming latencies in single trials. Said proportional costs were calculated for each participant.

Proportional switch costs were significantly higher in HD patients (6.2%) than healthy controls (1.7%) [F_1 (1, 26) = 5.07, $p < .05$, $\eta^2 = .17$; F_2 (1, 14) = 5.44, $p = .05$, $\eta^2 = .28$]. Additionally, a t-test revealed that this cost was not significantly different from zero in healthy controls [t (13) = 1.19, $p = .26$] while in HD patients it was significantly different from zero [t (11) = 4.83, $p = .01$]. Mixing costs were not significantly different between the two groups of participants [healthy control: 4.7%, HD patients: 4.0% [F_1 (1, 26) = .04, $p = .84$; F_2 (1, 14) = .01, $p = .94$].

Accuracy. The results revealed a significant main effect of type of trial [F_1 (2, 48) = 3.11, $p = .05$, $\eta^2 = .11$; F_2 (2, 28) = 4.91, $p < .05$, $\eta^2 = .26$], and post-hoc analyses demonstrated that

accuracy was lower for switch (97.5%) and repeat trials (97.6%) than single trials (99.0%, $p < .05$). Huntington's disease patients were also significantly less accurate (96.3%) compared to healthy controls (99.8%) [$F_1(1, 24) = 6.25, p = .02, \eta^2 = .21$; $F_2(1, 14) = 4.91, p < .05, \eta^2 = .17$].

INSERT FIGURE 1 ABOUT HERE

4.2. Stroop task

Naming latencies. The by-participants ANOVA showed a significant main effect of language context [$F_1(1, 24) = 17.55, p < .001, \eta^2 = .42$], with slower naming latencies in the within-language (863 ms) than the between-languages condition (821 ms) (Figure 2). However, the main effect of language context was not significant in the by-items ANOVA [$F_2(1, 10) = .55, p = .48$].

The main effect of type of trial was also significant [$F_1(2, 48) = 44.22, p < .001, \eta^2 = .65$; $F_2(2, 20) = 24.54, p < .001, \eta^2 = .71$]. Post-hoc analysis suggested that naming latencies were significantly different between the neutral (798 ms) and the incongruent conditions (920 ms, $p < .001$) but not between the neutral and the congruent conditions (807 ms) ($p = .36$).

The main effect of group was also significant [$F_1(1, 24) = 6.14, p < .05, \eta^2 = .20$; $F_2(2, 20) = 12.65, p < .01, \eta^2 = .56$], indicating that HD patients were slower overall (921 ms) than healthy controls (762 ms).

However, the interaction between the main effects of language context and type of trial was not significant [$F_1(2, 48) = .44, p = .65$; $F_2(2, 20) = .03, p = .97$] indicating that the within- and between-language versions were not different in terms of Stroop effects.

Accuracy. The main effect of type of trial was significant [$F_1(2, 48) = 4.81, p < .05, \eta^2 = .17$; $F_2(2, 20) = 6.27, p < .01, \eta^2 = .38$]. Post-hoc analysis suggested that accuracy was significantly lower in the incongruent (99.2%) than the neutral condition (99.8%), but no difference in accuracy was found between the neutral and congruent (99.9%, $p = .37$) conditions. No other main effect or interaction resulted significant in the analysis, suggesting that two groups of participants performed at the same level (HD patients = 99.4%, controls = 99.8%; $F_1(1, 24) =$

2.44, $p = .14$; $F_2(1, 10) = 1.37$, $p = .27$) and with no difference between the two versions of the task.

INSERT FIGURE 2 ABOUT HERE

5. Discussion and conclusion

We studied how two components of BLC (language activation/inhibition and cross-language interference) are impaired by a disease that affects the integrity of the striatum (HD). The results revealed a clear dissociation between these two mechanisms: while language activation/inhibition is affected by the pathology, presumably due to dysfunctions in the striatum, cross-language interference is not.

To test language activation/inhibition, we explored the modulation of preparation time for selecting the target language in a language switching task. The measure used for this BLC mechanism was the language switch cost; specifically, the difference in naming latencies between switch and repeat trials in a mixed-language condition. This language switch cost is often interpreted as resulting from the residual activation of the previous language, which interferes with lexical access in the current language (Philipp, Gade, & Koch, 2007) or reactive inhibition of the non-target language (Green, 1998). Importantly, this cost is reduced or even disappears when bilinguals have enough time to prepare the new naming language, as seen when they are given a longer preparation time (longer CSIs). We hypothesised that, since language activation and selection involves basal ganglia activity and more specifically striatum (Abutalebi & Green, 2008, 2016; Luk et al., 2011), patients with dysfunctions in these subcortical areas would not benefit from longer preparation times. Indeed, HD patients did not benefit from preparation time, suffering the same switch cost in both short or long CSI conditions, unlike healthy controls in which such cost was reduced as CSI became longer (Costa & Santesteban, 2004; Fink & Goldrick, 2015; Ma et al., 2016; Mosca & Clahsen, 2016; Verhoef et al., 2009).

Interestingly, however, in terms of switch costs, HD patients did not seem to be impaired in the language switching task with short CSIs since they showed comparable performance to healthy controls. This result contrasts with findings from a previous study in which bilingual PD patients experienced greater costs than healthy controls when switching between languages with short CSIs, suggesting that basal ganglia play a role in language inhibition (or “reactive control” according other EC models such as Braver, 2012). These contrasting observations are likely due to the fact that most of the patients tested in our study were in a pre-symptomatic stage or very early stage of the disease, and consequently only subtle changes in cognitive impairment can be detected. Pre-symptomatic HD patients are usually less impaired than those who are in the symptomatic stage of the disease for EC, and their deficits can be isolated to specific tasks (De Diego-Balaguer et al., 2008) or EC processes (e.g., Maurage et al., 2017; Tabrizi et al., 2013; You et al., 2014; for a review see Papp, Kaplan, & Snyder, 2011). In our sample, symptomatic patients were different from pre-symptomatic patients solely in the manifestation of motor impairments; we therefore assume that they were equivalent in terms of EC deficits.

Our results also speak to the difference between reactive and proactive control in HD. As described above, proactive control was measured by mixing costs, defined as the difference in naming latencies between repeat trials in the mixed language condition and single trials in the blocked language condition. Proactive control is a flexible control system needed to actively maintain goal-relevant information in a sustained manner (Cattaneo et al., 2015; Ma et al., 2016). Bilinguals use this type of control to actively maintain the two languages and monitor them during the task. We did not find any impairment of this mechanism due to the disease, and no difference was found in the magnitude of mixing costs between the HD patients and controls. Moreover, with longer CSIs, the magnitude of the mixing cost decreased in the same way as in healthy controls, suggesting that proactive language control is not impaired and is a sustained mechanism of BLC. Therefore, the dissociation between the impaired reactive control and the spared proactive one suggests that these controls are two distinct mechanisms dependent on qualitatively dissimilar processes, as Braver suggests for the EC system (2012; 2003). This result is interesting because it provides evidence for a specificity of mechanisms to subcortical structures such as the

striatum within the BLC network, as suggested in Abutalebi and Green's (2016) neural adaptation hypothesis. According to this revised version of the BLC network, these structures play a role in language selection (activation and inhibition) in conversation contexts that are more demanding because bilinguals switch back and forth continuously (dual-language) (see also Yan et al., 2018). These structures activate and inhibit languages in cooperation with the frontal areas for conflict resolution and parietal areas for maintaining language representations (see also Abutalebi & Green, 2008a; Branzi, Della Rosa, Canini, Costa, & Abutalebi, 2016; Seo, Stocco, & Prat, 2018; Zou, Ding, Abutalebi, Shu, & Peng, 2012). Alternatively, and specifically in relation to EC deficits in HD patients, it has been proposed that longer switch costs are related to response suppression at selection level (for a general account on response selection on HD patients see Lawrence, Sahakian, & Robbins, 1998). For instance, Aron et al. (2003) demonstrated that HD patients were greatly impaired compared to healthy controls in repeated response relative to alternated response in task switching. The researchers argued that excessive inhibition applied to the response that had just been performed impaired the suppression of the response and had a negative carry-over effect on the selection of the new response. Although we do not have measures of the brain damage of our patients, we speculate that our results may be interpreted similarly as due to dysfunction of basal ganglia in HD: the inhibition applied to the language used just before a switch was so excessive that it persisted even with longer preparation times.

To study the mechanisms of language interference, we used a within- and a between-language version of the Stroop task. Although patients were overall slower than healthy controls, they did not suffer more semantic interference in the incongruent condition. In fact, the overall pattern of results in both conditions were very similar for HD patients and healthy controls. This observation contrasts with our predictions. One potential explanation of why HD patients do not seem to have deficits in managing cross-language interference may be because our task only involved production in one language. It is then possible that the striatum is mostly involved in resolving cross-language interference in conditions where the two languages are both potential candidates to be produced as in the case of the language switching taskⁱ.

To conclude, our results would suggest that the striatum is a crucial structure in certain mechanisms involved in BLC. In particular, seeing that HD patients exhibit dysfunction in the striatum, it appears that said structure participates in the mechanisms that control language activation/inhibition. What is less clear is the involvement of the striatum in cross-language interference resolution, at least when only one language is used for production.

However, it must be acknowledged that the current study did not include neuroimaging data that would make it possible to establish a direct correlation between the damage of the striatum and language switching deficits in patients. Our hypothesis was based on the findings from other studies which reveal that the subcortical structures are affected since the start of the pre-symptomatic stage of HD and they consistently indicated that striatum is the one most damaged structures in pre-symptomatic patients (for volumetric reductions see Georgiou-Karistianis et al., 2013; for alterations in diffusion indices of basal ganglia see Rosas et al., 2006; Vandenberghe et al., 2009). Future neuroimaging studies would help to make a more direct connection between how damage to this structure alters bilinguals' abilities to control their two languages.

¹ Moreover, the activation of the left caudate in monolinguals performing the Stroop task has been reported (Ali et al., 2010), and this result has been interpreted as reflecting the suppression of verbal interference. It may be the case that in HD patients, the degeneration of the striatum affects performance in the Stroop task as a deficit of verbal response inhibition independent of the language used. Indeed, this relationship would explain why this task is sometimes sensitive to detecting early cognitive deficits in monolingual patients with HD (Baake et al., 2017; Montoya, Price, Menear, & Lepage, 2006; Peavy et al., 2010; Peinemann et al., 2005; Stout et al., 2011; Tabrizi et al., 2013).

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Figure 1. Naming latencies, proportional switch and mixing costs for the language switching task as a function of CSI and group of participants

Figure 2. Naming latencies, proportional semantic and identity effects for the Stroop tasks as a function of language context and group of participants

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Appendix

BILINGUALISM QUESTIONNAIRE

1. Language acquisition and usage

a. Current city of residence

If different from the place of birth, specify when (year) you moved to your current residence

b. Father's place of birth/Language(s) spoken

c. Mother's place of birth/Language(s) spoken

d. Catalan – Age of acquisition

At what age were you exposed to Catalan?

At what age did you begin to use (speak) Catalan?

e. Spanish – Age of acquisition

At what age were you exposed to Spanish?

At what age did you begin to use (speak) Spanish?

f. Where did you learn Catalan? At home At school Other

g. Where did you learn Spanish? At home At school Other

h. Specify in which language (or languages) you are used to speak to:

Father:	Mother:	Partner:
Sisters/Brothers:	Children:	

i. Do you speak any other languages besides Catalan and Spanish?

j. What was the age of acquisition for this/these language(s)?

2. Language proficiency

Rate your proficiency in the following aspects of language within Catalan, Spanish, and any other languages you speak:

a. Speaking

Catalan	Perfect	Good	Regular	Poor
Spanish	Perfect	Good	Regular	Poor
L3	Perfect	Good	Regular	Poor
L4	Perfect	Good	Regular	Poor

b. Comprehension

Catalan	Perfect	Good	Regular	Poor
Spanish	Perfect	Good	Regular	Poor
L3	Perfect	Good	Regular	Poor
L4	Perfect	Good	Regular	Poor

c. Reading

Catalan	Perfect	Good	Regular	Poor
Spanish	Perfect	Good	Regular	Poor
L3	Perfect	Good	Regular	Poor
L4	Perfect	Good	Regular	Poor

d. Writing

Catalan	Perfect	Good	Regular	Poor
Spanish	Perfect	Good	Regular	Poor
L3	Perfect	Good	Regular	Poor
L4	Perfect	Good	Regular	Poor

e. Which language do you consider to be your dominant language?

Catalan ☐ Spanish ☐

f. In which language do you feel more comfortable speaking?

Catalan ☐ Spanish ☐ Both ☐

g. Are you able to distinguish speakers' regional accents?

Catalan	No	In some cases	Yes, always
Spanish	No	In some cases	Yes, always

3. Frequency of language use and dominance

In answering the questions below, specify the frequency with which you used Spanish and/or Catalan during the different periods of your life and according to the contexts described below. Use the following scale for your responses:

0	Only Spanish
1	Mostly Spanish, rarely Catalan
2	Frequently Spanish with $\frac{1}{4}$ of the time in Catalan
3	Same time for Spanish and Catalan
4	Frequently Catalan with $\frac{1}{4}$ of the time in Spanish
5	Mostly Catalan, rarely Spanish
6	Only Catalan

a. As a child, before formal schooling?

<i>only Spanish</i>				<i>only Catalan</i>		
0	1	2	3	4	5	6

b. As an adolescent, from 4-5 to 12 years old?

	<i>only Spanish</i>				<i>only Catalan</i>		
At school	0	1	2	3	4	5	6
At home	0	1	2	3	4	5	6
With friends	0	1	2	3	4	5	6

c. As a teenager, from 13 to 16 years old?

	<i>only Spanish</i>				<i>only Catalan</i>		
At school	0	1	2	3	4	5	6
At home	0	1	2	3	4	5	6
With friends	0	1	2	3	4	5	6

d. As a young adult, from 17 to 22 years old?

	<i>only Spanish</i>				<i>only Catalan</i>		
At school/university	0	1	2	3	4	5	6
At home	0	1	2	3	4	5	6
With friends	0	1	2	3	4	5	6

e. In adulthood?

	<i>only Spanish</i>				<i>only Catalan</i>		
At work	0	1	2	3	4	5	6
At home	0	1	2	3	4	5	6
With friends	0	1	2	3	4	5	6