28 Primary Progressive Aphasias in Bilinguals and Multilinguals

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1. Introduction

Primary progressive aphasia (PPA) is the result of neurodegeneration affecting language abilities that continue to decline as the disease progresses. There are three main variants of PPA: non-fluent, semantic, and logopenic. Deficits may occur in different areas of language, such as lexical retrieval, auditory comprehension, syntactic structure, processing morphological components, and repetition abilities. However, the impact on language is not comparable across all individuals with PPA; rather it differs for each of the different variants based on the underlying pattern of neural change.

In bilinguals or multilinguals with PPA, the language decline has an added layer of complexity. Decline may occur across the different languages in parallel, or differentially, and a number of factors may affect the pattern of decline. Recognizing the factors that most affect language decline in bilinguals and multilinguals with PPA, along with identifying the neural changes occurring in the brain, can increase our understanding of language organization in the bilingual or multilingual brain. It should be noted that language decline is not the only decline associated with PPA, as changes in cognition and behaviour have also been observed, particularly in the later stages (e.g. Rosen et al. 2006). However, language is the most salient decline in PPA so we focus on language in this chapter.

We analysed 13 case-studies of bilinguals and multilinguals with PPA published to date, which included all three variants of PPA, and found that language decline across languages within an individual can be differential and/or parallel. We discuss that in the cases of differential language decline, the factor that appears to most strongly affect the pattern of decline is the order of acquisition, in that the first-acquired language was better spared than any later-acquired languages. Other factors, such as proficiency,

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recency of use, manner of acquisition, age of acquisition, and language typology (which includes aspects like language distance) did not appear to have a strong effect on either differential or parallel decline. In most cases, aspects of parallel and differential decline were observed across different language domains. However, even in those domains where differential decline was observed, there was a shift towards parallel decline as the PPA progressed.

We will discuss language decline in bilinguals and multilinguals with PPA in relation to language decline in bilinguals with another degenerative disease (Alzheimer's disease) as well as in relation to sudden-onset aphasia in bilinguals as a result of a cerebrovascular accident (CVA). Due to the degenerative nature of PPA, carefully analysing language decline in bilinguals and multilinguals with PPA can add to our knowledge of bilingual and multilingual language organization in the brain and the effect of neural impairment on language.

For ease of reading, *bilinguals with PPA* will refer to either bilinguals or multilinguals for the remainder of this chapter, unless specifically stated otherwise. We begin the next section with a detailed explanation about PPA and its three main variants, following which we discuss different hypotheses of language organization in the bilingual brain, at the neural level. This will then lead us to an in-depth discussion about bilinguals with PPA, patterns of language decline, important factors affecting this decline and how this supports or conflicts with existing models of language organization in the bilingual brain.

1.1. Primary Progressive Aphasia

PPA is a subtype of dementia with progressive decline in language abilities over time relative to cognitive abilities that decline late in the disease. In contrast to sudden-onset aphasia, in PPA there is no defined lesion, nor is there diffuse cortical atrophy. Rather there is progressive atrophy initially to a somewhat confined region of the brain (Gorno-Tempini et al. 2008) as indicated, for example, by atrophy on magnetic resonance imaging (MRI) scans (Gorno-Tempini et al. 2004) or hypoperfusion on positron emission tomography (PET) scans (Sinnatamby et al. 1996). Even within these sites of cortical atrophy in patients with PPA, neuronal destruction is never fully complete, and the remaining neurons continue to participate in language function, but with altered patterns of neural network connectivity (Mesulam et al. 2014).

Different subtypes of PPA have been associated with volume loss in specific cortical regions. Despite differences in regional volume loss amongst the PPA subtypes, cortical atrophy is mostly confined to the left hemisphere in the non-fluent and logopenic variants (e.g. Gorno-Tempini et al. 2011; Mesulam 2007). In the semantic variant, by contrast, atrophy is usually seen in both hemispheres, yet in most cases, atrophy is more pronounced in the left hemisphere than the right hemisphere (Mion et al. 2010; Rogalski et al. 2011). Besides left-dominant atrophy, the semantic variant can also present with right-dominant atrophy (Gorno-Tempini et al. 2004). In such cases, early semantic decline is often more pronounced in the non-verbal than verbal domain. The subtle language impairment in the early stages of the right-dominant variant grows stronger with progression of the disease along with increasing atrophy in the left hemisphere

(Binney et al. 2016; Vonk et al. in preparation). In this chapter, however, we focus on the classic, more common left-dominant variant.

Language decline in PPA occurs as a result of these neuroanatomical changes of progressive cortical atrophy. Each of the three subtypes of PPA (non-fluent, semantic, and logopenic variants) presents with differential patterns of atrophy that result in different clinical symptoms and patterns of language decline.

In the non-fluent variant, cortical atrophy and hypometabolism are found in the left inferior frontoinsular area (Gorno-Tempini et al. 2011), although hypoperfusion has also been documented in the left superior and middle temporal areas secondary to the inferior frontal area (Grossman 2010). Damage to these areas results in impaired speech output, characterized by simple sentences, poor syntactic structure, and speech sound errors. Such speech is described as telegraphic since it omits essential morphological components (Gorno-Tempini et al. 2004). However, people with this variant of PPA have relatively preserved auditory comprehension for the majority of the duration of this decline (Hodges and Patterson 1996; Thompson et al. 1997).

In the semantic variant, atrophy is focused around the anterior and inferolateral temporal areas. As mentioned earlier in this section, atrophy in this variant is generally bilateral (Mion et al. 2010). However, it tends to be greater on the left than on the right, especially in the early stages of the disease (Rogalski et al. 2011). This pattern of atrophy results in severe deficits in confrontation naming (Hodges and Patterson 1996) due to deficits in semantic memory. Speech output appears fluent and is well-articulated but the content is empty, and errors include semantic paraphasias, circumlocutions, and non-specific names (e.g. Blair et al. 2007). Given that the deficit is found in semantic memory, language impairments are observed both in verbal expression as well as comprehension and recognition of single words and objects (Mesulam 2003).

Logopenic PPA shows atrophy in the left posterior temporoparietal region or hypoperfusion in the same region early in the disease and later in the anterior temporal area (Gorno-Tempini et al. 2011). Symptoms include word-finding deficits, leading to frequent hesitations and pauses in speech. These hesitations make speech sound dysfluent, with increasing dysfluencies when content requires precision (Mesulam 2007; Wilson et al. 2010). Errors are characterized as phonemic paraphasias, decreased naming ability, and reduced repetition ability. Gorno-Tempini et al. (2011) further specified that in logopenic PPA there is an absence of frank agrammatism, differentiating it from the non-fluent variant.

This influence of brain atrophy on language impairment in the three variants is relatively consistent amongst monolinguals with PPA. Language impairment in bilinguals with PPA is more complex but can provide valuable information about how two or more languages are represented in the brain. We will now discuss a number of issues of bilingual brain organization which may help us better understand the patterns of language impairment in bilingual PPA. In Section 1.2, we will focus on bilingualism, rather than multilingualism, in the brain because that is the focus of the existing literature to date. Currently, differences between bilingualism and multilingualism at the neurological level are still unclear both generally and specifically in PPA.

1.2. Bilingualism in the Brain

With the development of neuroimaging techniques in the last three decades, knowledge of bilingual neural organization in healthy children and adults has increased, as well as in clinical populations such as bilinguals with sudden-onset aphasia and dementia. Together, behavioural and neuroimaging studies have provided much evidence that at least some language systems are shared by the first language (L1) and second language (L2; e.g. Kroll and Tokowicz 2005; Perea et al. 2008). However, there is also neuroimaging evidence of structural or functional differences in brain regions for L1 and L2. For example, studies of healthy adult bilinguals have found shared processing regions in high proficiency bilinguals, but more limited overlap of processing regions in low proficiency bilinguals (e.g., Dehaene et al. 1997; Perani et al. 1998). Interestingly, the regions additionally activated for low-proficiency bilinguals were all in the right hemisphere: right middle temporal gyrus for L1, and right hippocampus, and superior parietal lobule for L2. Similarly, Kim et al. (1997) also showed differential activation for L1 and L2 when comparing early versus late age of acquisition, finding less overlap of regions in the left inferior frontal lobe during covert naming when L2 was acquired late.

Studies examining white matter pathways have also found differential effects in late bilinguals. Kuhl et al. (2016) examined late bilinguals immersed in the environment of the L2 compared with monolinguals. The study found white matter activation and diffusion to be bilaterally activated in bilinguals; in monolinguals activation is less bilateral. The bilingual group showed a correlation of duration and activation, such that those with longer immersion in the environment of the L2 exhibited less diffusion in white matter tracts (as measured by fractional anisotropy) in specific language pathways than those with shorter immersion in the environment of the L2.

Together, these studies suggest that factors such as level of proficiency, age of acquisition, and recency of use may alter the organization of language in the brain in healthy bilinguals. Nonetheless, the lack of consensus as to the neural representation of bilinguals' languages has resulted in the development of different models for language organization in the bilingual brain, based on factors related to individual profiles of bilinguals. Two of these well-supported but opposing models have dominated this field: the convergence hypothesis and the declarative-procedural model.

The convergence hypothesis (Green 2003) states that neurocognitive mechanisms of second language learning can improve with increased practice, thereby converging on L1 neural networks. Changes in proficiency are associated with shifts from more controlled to more automatic language processing (Abutalebi and Green 2007). In fact, rapid convergence of neural patterns has been shown both by functional magnetic resonance imaging (fMRI; Consonni et al. 2013; Golestani et al. 2006; Van de Putte et al. 2017) and event-related potential (ERP) data (Osterhout et al. 2006). However, this convergence may depend on linguistic variables other than (or as well as) proficiency, such as linguistic distance (Chen et al. 2007).

While current neuroimaging research is consistent with the position that the languages of bilinguals are at least partially represented in shared processing regions

(e.g. Abutalebi et al. 2001), the literature has not yet fully accounted for evidence of differential organization – such as when languages are affected differently by brain damage. However, most agree that conceptual representations in different languages share a common substrate when considering lexical-semantic processing (e.g. Crinion et al. 2006; Stilwell et al. 2016) and there is also evidence of common regions mediating syntax (Golestani et al. 2006; Ullman 2015).

An alternative way to explain these differential findings is the declarative-procedural model which characterizes variability in language impairment following neural damage (Paradis 2003; Ullman and Pierpont 2005). In addition to proficiency, this model uses age of acquisition (early vs. late) and manner of acquisition (i.e. context of L2 acquisition) to explain why a second language may be represented differently in the brain. The model hypothesizes that a second language acquired early may be neurally represented differently from a late-acquired second language and that an early-acquired L2 will be more similarly represented in the brain to L1 than a late-acquired L2 under certain learning contexts, independent of proficiency (Ullman 2015). Additionally, L2 acquired in the home will be acquired more with procedural memory, i.e. implicitly, whereas acquiring an L2 at school will be accomplished more explicitly, therefore incorporating more declarative memory.

The declarative-procedural model proposes that both L1 and L2 use declarative memory to store idiosyncratic lexical knowledge in different linguistic domains, including simple words and their meanings, irregular morphology, and syntactic complements (Ullman 2015). At the same time, syntactic and morphological processes in L1 are mediated by the procedural system typically responsible for other cognitive and motor skills (Paradis 2008; Ullman 2015). L2 acquisition of syntactic and morphological processes can rely on the procedural system when acquired early, but the declarative system plays a crucial role in the representation of grammar if the L2 is acquired late. The use of declarative and procedural memory may result in differential effects of brain damage in L1 and L2, depending on age of acquisition and manner of acquisition. In bilinguals, when L2 is acquired late, the declarative system will play an important role in the production of morphological and syntactic forms that rely on the procedural system in L1. As L2 proficiency increases, the use of the procedural system to process it will likely increase, especially if it is acquired implicitly (e.g. via immersion; Ullman 2015). When the system is damaged, early bilinguals with high proficiency in both languages may have comparable performance in L1 and L2 in morphological and syntactic tasks.

These two models – the convergence hypothesis and the declarative-procedural model – follow a clear pattern, in that differential effects may be seen in brain damage based on individual factors of bilinguals. However, each model attributes differences in bilingual language organization to different factors: while the convergence hypothesis emphasizes proficiency and use of each language, the declarative-procedural model emphasizes manner and age of acquisition. To test which model best fits the patterns of language decline in bilinguals with PPA to date, we ask which factors are important for language decline in bilinguals with PPA and how these relate to language loss in other similar populations – i.e. sudden-onset aphasia and dementia in bilinguals.

2. Bilingual and Multilingual PPA

While the characterization of PPA began 30 years ago, much of the literature has examined monolingual individuals with PPA, establishing the three distinct variants mentioned previously. Cases of bilinguals or multilinguals with PPA began to be examined around the same time the current PPA classification was developed (in the early 2000s). To date, we have identified 12 published studies (one an abstract) on bilingual or multilingual PPA. All of these publications examined either single cases or two individuals with similar variants of the disorder. Two papers examined two individuals, one looking at two bilingual participants with the semantic variant (Mendez et al. 2004) and the other examining one multilingual and one bilingual, also both characterized as having the semantic variant (Liu et al. 2012). However, we removed the second case in the Liu study from consideration in this chapter, as his behavioural data indicated more widespread cognitive deficits than PPA usually allows for, throwing doubt to his PPA diagnosis.

After eliminating this one participant, we were left with 12 published studies which included 13 different case studies of bilinguals and multilinguals with PPA. Five have the non-fluent variant, four have the semantic variant, and four have the logopenic variant. Diagnoses and variant classifications were determined with both behavioural data from a variety of language and cognitive tasks, as well as neuroimaging data from MRI, PET, single-photon emission computerized tomography (SPECT), and/or computerized tomography (CT) scans. These case studies were analysed for the following factors: PPA variant, site of cortical atrophy, languages acquired, age of acquisition, dominance and/or proficiency, language(s) of the environment, and patterns of language abilities and decline in each language. See Table 1 for details of demographic information for each case study.

The most striking factor that can be observed when looking across all cases is that regardless of variant, in no case was L2 better preserved than L1. In all cases, either L1 was better preserved than L2, or both languages declined in parallel. Cases that found L1 better preserved than L2 assessed language decline via a variety of subtests; these varied across studies. For non-fluent PPA, all five studies showed that L2 was never better preserved than L1 for object and action naming (Druks and Weekes 2013; Hernàndez et al. 2008; Larner 2012; Machado et al. 2010; Zanini et al. 2011), a finding also present in one case with the logopenic variant (Filley et al. 2006; Lind et al. 2017). Similarly, two cases of multilinguals with the semantic variant showed greater decline in L2 than L1, with L3 being almost completely lost, in word comprehension and naming (Mendez et al. 2004). There was also a noted difference between tasks of grammatical processing in one case, with L2 declining more rapidly than L1 (Zanini et al. 2011).

Those areas of language showing parallel decline included lexical-semantic knowledge and access, phonological knowledge, comprehension of complex commands, and reading and writing abilities (Devaughn et al. 2016; Druks and Weekes 2013; Filley et al. 2006; Hernàndez et al. 2008; Liu et al. 2012; Zanini et al. 2011). Grammatical knowledge was also found to decline in parallel for one case (Druks and Weekes 2013). In the six cases tested at multiple time points, all deficits eventually declined in parallel as the disease progressed to the later stages. For example, Druks and

Table 1 Demographic information for 13 published case studies on bilinguals or multilinguals with PPA.

	PPA variant	*Cortical atrophy*	*Languages*	*Age of L2 Acquisition*	*Dominant language / proficiency*	*Language of environment*	*Manner of acquisition of L2 (and/or L3)*	*Most preserved language overall*
Mendez et al. (2004) – P1	Semantic	Left anterior temporal	L1-English L2-Spanish L3-German	N/A	Proficient in L1 and L2 L3[a]	L1	N/A	L1
Mendez et al. (2004) – P2	Semantic	Left anter ior temporal	L1-Spanish L2-English L3-Polish	N/A	Proficient in L1 and L2	N/A	N/A	L1
Filley et al. (2006)	Logopenic	Left temporal and parietal	L1-Chinese L2-English	8 years	L2 dominant	L2	Formal, school context	L1 = L2
Hernàndez et al. (2008)	Non-fluent	Perisylvian cortex and hippocampus; right prefrontal region	L1-Spanish L2-Catalan	Before 4 years	Balanced, proficient	L1 and L2	Formal, school context	L1
Machado et al. (2010)	Non-fluent	Left temporal, anterior cingula, and dorsolateral frontal	L1-Portuguese L2-French	4 years, then again as an adult	Balanced	L1	Acquired in the home	L1
Zanini et al. (2011)	Non-fluent	Left frontal, parietal and temporal	L1-Friulian L2-Italian	6 years	Balanced	L1 and L2	Acquired in the home	L1
Druks and Weekes (2013)	Non-fluent	Left perisylvian atrophy; intact medial temporal and posterior structures	L1-Hungarian L2-English	14 years	L2 dominant	L2	Formal, school context	L1 = L2
Kambanaros and Grohmann (2012)	Logopenic	Left temporal parietal	L1-Cypriot L2-Greek L3-English L4-Czech	L2 & L3 –Early exposure L4 – Late exposure	Highly proficient L1, L2, L3, L4	Mainly L1	Acquired in the home; and formal, school context	L1 (informally evaluated)

Liu et al. (2012)	Semantic	Left inferior temporal gyrus	L1-Taiwanese L2-Japanese L3-Mandarin Chinese	L2 – 7 years L3 – 11 years	Proficient in L1, L2, L3	L1 and L3	Formal, school context	L1 = L2
Larner (2012)	Non-fluent	Left insular and anterior left temporal lobe	L1-Welsh L2-English	6 years[a]	N/A	L2	Formal, school context	L1
Meyer et al. (2015)	Logopenic	Left temporo-parietal	L1-Norwegian L2-English	7 years	L2 dominant	L2	Immersion in environment	L1 = L2
DeVaughn et al. (2016)	Semantic	N/A	L1-English L2-German	N/A	Balanced	L1	N/A	L1 = L2
Lind et al. (2017)	Logopenic[a]	Bilateral, large areas of the brain; not specified	L1-English L2-Norwegian	31 years	L1 dominant	L1 at home; L1 and L2 at work	Immersion in environment	L1

[a] indicates that the information has been inferred from the article, rather than explicitly stated by the authors.
N/A – information not available, even from inference.

Weekes (2013) found that most language tasks declined in parallel. However, in the two linguistic tasks where L1 was better spared than L2 at the first testing point two years post-onset (namely object and action naming), when retested one year later, L1 and L2 showed parallel decline in these two tasks along with the other language tasks. Similarly, Machado et al. (2010) found that L2 declined more drastically than L1 in their participant when tested two years post-onset, but within a year the L2 and L1 were similarly impaired. From these cases, we can see that language decline appears to become parallel over time in PPA, but how this decline progresses and under what circumstances the decline is parallel from the time of diagnosis, or whether the decline is differential from the time of diagnosis before changing to parallel at a later stage, remains unclear.

2.1. Factors Impacting Language Decline

While the literature on bilinguals with PPA seems to show that variant type has little effect on whether there is parallel decline or better L1 preservation, it is important to look at other factors that may impact how the two languages are organized in bilinguals, potentially resulting in differential decline. Here we ask whether age of acquisition, manner of acquisition, proficiency level, or recency of use (related to language of environment) impact better preservation of one language or if the languages decline in parallel. Due to the limited literature, it would be reasonable to compare the PPA literature with two better-studied fields in this regard: Alzheimer's disease (AD) and sudden-onset aphasia.

In bilinguals with AD, many researchers have documented better-preserved language abilities in L1 than in L2 (e.g. Ardila et al. 2008; Meguro et al. 2003; Mendez et al. 1999; Stilwell et al. 2016), although others have observed parallel deterioration between L1 and L2, especially as the disease progresses (e.g. Costa et al. 2012; Gómez-Ruiz et al. 2012; Manchon et al. 2015). In addition, language dominance rather than order of acquisition has been suggested to affect language performance in AD in some cases (see Stilwell et al. 2016, for a review), but closer analysis of the literature shows that the two studies they cite to support this hypothesis are not strong support at all. In one study by Gómez-Ruiz et al. (2012), the participants were balanced, early bilinguals of Catalan (L1) and Spanish (L2), and showed parallel deterioration overall. The only place the authors found a significant difference between L2 and L1 was in the one subtest related to reading and writing – namely verbal letter fluency (i.e. naming as many items as possible beginning with a given letter). In this population of bilinguals, the L2 (Spanish) was the language of schooling and the first (or in a few specific cases the only) language acquired for reading and/or writing. Clearly this single subtest is not good support for suggesting that language dominance may affect language performance more than order of acquisition in AD, since for literacy, Spanish was actually the first-acquired language.

In the second study by Gollan et al. (2011), two groups of participants were tested – L1 dominant and L2 dominant (premorbid dominance). Overall, the L2 dominant participants had relatively mild dementia compared to the L1 dominant group, where the dementia was more severe. The results showed that for the L1 dominant group, L2 was better spared than L1, but the results did not even approach significance. Also, the

testing measure used (the Boston naming test; Kaplan et al. 1983), was noted by Gollan et al. to be problematic when comparing across English and Spanish, since the test was developed for English speakers residing in the US. The use of this test together with the more severe dementia, as well as the lack of significance, we would argue, is not strong evidence that L2 is sometimes spared more than L1 in AD. Therefore, we conclude, the two main patterns of language decline in bilinguals with AD are similar to those in bilinguals with PPA.

In sudden-onset aphasia, by contrast, several different parallel and non-parallel patterns have emerged to characterize language of bilinguals (Paradis 2001). Factors such as age of acquisition, proficiency, language dominance, and recency of use have all been found to contribute to language impairment (and recovery) post-stroke (e.g. Faroqi-Shah et al. 2010; Goral et al. 2012; Lorenzen and Murray 2008). The 13 PPA cases reviewed here have shown no distinct pattern as to individual factors. The only certainty is that at no point is L2 better preserved than L1, seemingly regardless of age of acquisition, manner of acquisition, proficiency level, or recency of use. For example, amongst the 13 cases, there was a range of ages of acquisition. In three cases, L2 was acquired very early in childhood (Hernàndez et al. 2008; Kambanaros and Grohmann 2012; Machado et al. 2010). A further five cases were characterized as having acquired L2 from age six up to puberty (Filley et al. 2006; Larner 2012; Liu et al. 2012; Meyer et al. 2015; Zanini et al. 2011). Only two cases discussed L2 acquisition post-puberty: Druks and Weekes' (2013) participant was age 14 and Lind et al.'s (2017) participant was age 31. The cases of DeVaughn et al. (2016) and Mendez et al. (2004) did not provide information about age of acquisition. For multilinguals, Liu et al. (2012) described a case where L3 was acquired later than L2, but still acquired pre-puberty, and Kambanaros and Grohmann (2012) described a case where L3 was acquired in early childhood and L4 acquired in adulthood. The range of ages of acquisition amongst these studies clearly demonstrates that, thus far, age of acquisition for a second language or later learned language is not a major factor for language decline in PPA.

As discussed in Section 1.2, the declarative-procedural model also pertains to how the manner of acquisition can impact language retention, with use of declarative or procedural memory to acquire different linguistic domains for L1 and L2. This use of different memory systems is hypothesized to explain differences found in language recovery from sudden-onset aphasia. Yet across all bilingual PPA cases examined, there was a range of manner of acquisition amongst participants, with some learning languages at home from a young age (Machado et al. 2010; Zanini et al. 2011), some learning in a formal, school context (Druks and Weekes 2013; Filley et al. 2006; Hernàndez et al. 2008; Larner 2012; Liu et al. 2012), some through immersion in the environment of the L2 (Lind et al. 2017; Meyer et al. 2015), and one multilingual case whose languages were acquired both at home from a young age and in a formal school context (Kambanaros and Grohmann 2012). As with age of acquisition, manner of acquisition was not observed to impact the overall outcomes of language decline in bilinguals and multilinguals with PPA.

Both proficiency level and recency of use have also been shown to be important factors for brain organization in healthy bilinguals, and as discussed in Section 1.2, the convergence hypothesis emphasizes them both. For proficiency, neuroimaging studies

have shown that level of L2 proficiency can change the neural representation of a second language in the brain in healthy bilingual adults (Dehaene et al. 1997; Kuhl et al. 2016; Perani et al. 1998) resulting in relatively differential neural representations for low proficiency L2 users and relatively converged neural representations for high proficiency L2 users.

However, no studies to date in the bilingual PPA literature show that proficiency has a strong effect on the pattern of language decline. Of the 13 cases of bilingual and multilingual PPA, all 13 were found to be proficient in L2, with eight being described as balanced bilinguals or highly proficient in both languages (Devaughn et al. 2016; Hernàndez et al. 2008; Kambanaros and Grohmann 2012; Liu et al. 2012; Machado et al. 2010; both participants studied by Mendez et al. 2004; Zanini et al. 2011), three with L2 dominance (Druks and Weekes 2013; Filley et al. 2006; Meyer et al. 2015), and one with L1 dominance (Lind et al. 2017). Even when conditions seem most likely that L1 and L2 will be neurally similar, in the cases where early bilinguals were balanced in proficiency and used both languages daily, decline was not necessarily parallel as the bilingual aphasia literature would lead one to expect. For example, Hernàndez et al. (2008) and Zanini et al. (2011) described early, balanced bilinguals who used both languages daily, yet for both cases L1 was better preserved than L2, either in noun and verb naming (Hernàndez et al. 2008) or a variety of language tasks (Zanini et al. 2011).

The only hint that proficiency may affect language decline in PPA arises when we contrast the non-L1 languages in two cases of multilinguals. Mendez et al. (2004) describe one case of a multilingual whose L1 and L2 had been highly proficient, but L3 was less proficient. In this case, L3 appeared to be completely lost when he was tested, whereas L1 and L2 declined – but L1 was better spared relative to L2. Compare this with the study by Kambanaros and Grohmann (2012) of a multilingual who was highly proficient in all four of his languages, was tested in all but the L1, and showed language processing in non-L1 languages declining in parallel across all language domains: morphology, phonology, lexical semantics, comprehension, and repetition (Kambanaros and Grohmann 2012). It is important to note that even in this case, the authors described the L1 as being better preserved than the other three languages; however that conclusion was determined only informally since their testing methodology – the bilingual aphasia test (Paradis 1987) – was not available in the participant's native language, Cypriot Greek (Kambanaros and Grohmann 2012). Based on these two cases, proficiency may be a factor in language decline in multilinguals with PPA, but order of acquisition still seems to be the strongest factor, such that L1 is better preserved even when other languages are as proficient as the L1.

Recency of use is a term used in the literature in relation to recovery from sudden-onset aphasia in bilinguals, where the most recent language is that being used at the time of the CVA (e.g. Faroqi-Shah et al. 2010; Goral et al. 2012). However, in the 13 cases of PPA in bilinguals and multilinguals that we reviewed, we did not find recency of use to be a critical factor affecting patterns of language decline. In fact, in all 13 cases, the L2 was used daily at least until the onset of PPA. In two cases, the participants were dominant in their L2 and rarely used their L1 (Druks and Weekes 2013; Filley et al. 2006). Even so, despite recency of use and immersion in L2, Filley et al. (2006) described L1 and L2 declining equally in naming after diagnosis of PPA, with more phonemic

paraphasias noted in connected speech in the L2 – the most recently used language. Similarly, Druks and Weekes (2013) showed parallel decline in most language tasks, except for object and action naming, which declined more in the L2 – the most recently used language.

However, in both Filley et al. (2006) and Druks and Weekes (2013), it is difficult to tease apart the effects of premorbid proficiency from the effects of recency of use. The one case study that does differentiate between these factors is that of Larner (2012) who examined a case of a Welsh-English bilingual. Though Welsh was the participant's native language, she was highly proficient in English and primarily spoke English in the home at the time of her PPA diagnosis, at age 78. Again, despite being proficient in both L1 and L2, the most recently used language – English (her L2) – declined more than the less recently used L1.

Therefore, proficiency level and recency of use were not found to be major factors influencing language decline patterns between L1 and L2 in bilinguals and multilinguals with PPA. Filley et al. (2006) argued that their findings may have been due to the language distance of the specific language pair in the bilingual (Chinese and English), and this may have resulted in less overlap in the neural organization of the two languages than in two more similar languages due to a mismatch in the tasks administered. However, in the studies we reviewed, several different language combinations were examined: Hungarian-English (Druks and Weekes 2013), English-German (Devaughn et al. 2016), Spanish-Catalan (Hernàndez et al. 2008), Portuguese-French (Machado et al. 2010), Friulian-Italian (Zanini et al. 2011), Norwegian and English (Lind et al. 2017; Meyer et al. 2015), Cypriot-Greek-English-Czech (Kambanaros and Grohmann 2012), Taiwanese-Japanese-Mandarin (Liu et al. 2012), Welsh-English (Larner 2012), English-Spanish-German, and Spanish-English-Polish (Mendez et al. 2004). Again, regardless of the linguistic distance in any given language combination, the L2 was never better preserved than the L1, and most languages declined in parallel for most language tasks. The difficulty with characterizing PPA is that it does not follow the typical patterns, and does not appear to be influenced by the same factors, as sudden-onset aphasia in bilinguals; early age of acquisition, proficiency, and recency of use have been shown to have little effect on PPA decline. Therefore, the focus should be more on the degenerative nature of the PPA, since the patterns of language decline are more consistent with those seen in AD.

2.2. Neurological Basis of Language Decline in PPA

The bilingual PPA literature partially supports the theory of a shared neural substrate for two or more languages, in that atrophy to certain cerebral regions in the brain results in decline in both languages, albeit not necessarily in parallel. Furthermore, the patterns of decline are consistent, despite individual bilingual factors and different language typologies. What remains unknown is why brain atrophy in PPA affects language differently from sudden-onset aphasia that occurs after a CVA (stroke), when they occur in the same brain regions.

In stroke, lesions of brain damage are localized around major arteries, and as such, certain areas of the cortex are typically spared from damage, in particular, the temporal pole.

PPA, by contrast, is caused by protein abnormalities within a neuron. In healthy neurons, proteins, such as tau proteins, stabilize the shape of the cell. In neurodegenerative diseases, the pathway for these and other proteins become tangled, resulting in the inability to maintain the structure of the cell, and thus individual neurons start to degenerate (Iqbal et al. 2010). Neurodegeneration of cells in PPA has been linked to tauopathy, TDP-43 proteinopathy (for the non-fluent and semantic variants) and Alzheimer's pathology (for the logopenic variant) and these proteins are unable to sustain the vitality of neurons in a certain region (Grossman 2010; Santos-Santos et al. 2018). Due to the death of individual cells in PPA, damage is not limited to areas surrounding major cerebral arteries. Instead, each variant has a typical epicentre for atrophy where it starts and remains – but as the disease progresses, atrophy spreads to other regions, resulting in extensive degeneration.

The other crucial difference between sudden-onset aphasia and PPA is the type of neural damage. In a stroke, grey and contiguous white matter is suddenly destroyed. Contrastingly, neurodegenerative diseases – such as those resulting in PPA – can target specific layers and regions of the cerebral cortex (Mesulam et al. 2014). Despite localized regions of cortical atrophy in PPA, neuronal death in any given region is not complete. In the early stages of the disease, only some neurons have degenerated, and the remaining neurons continue to function for language tasks. Due to the gradual nature of neuronal loss, the existing neurons can reorganize, at least to an extent, and retain some language function until the end-stage of the disease process.

In a stroke, however, neural injury results in more abrupt damage, with sudden and sometimes complete loss of language function. As a result of this sudden loss, the brain cannot gradually reorganize to retain certain language processes, but instead may rely on alternative pathways to regain some function. These differences between gradual decline and sudden loss lead to subtle (or not-so-subtle) differences in language function between the two languages of a bilingual, depending on the aetiology of language impairment.

Another difference between the neural atrophy of PPA and the cerebrovascular lesions of stroke is the differential impairment of white matter pathways. While white matter can be impaired in PPA, typically this impairment is not as severe as that in patients after a stroke. An example of this is seen in logopenic PPA which, as mentioned, is characterized primarily by deterioration of cells in temporoparietal cortical regions. While a vascular lesion in the same neural regions should indicate Wernicke's aphasia, with poor comprehension and fluent, but empty speech, temporoparietal atrophy in logopenic PPA does not result in impaired comprehension (For a more complete description of Wernicke's aphasia, see Kemmerer 2014). Mesulam et al. (2015) hypothesized that severe word comprehension deficits might only occur in PPA if white matter pathways are damaged, disconnecting posterior temporal regions from the anterior temporal lobe. The difference between atrophy and cerebrovascular lesions in similar areas demonstrates that the neural substrates of word and sentence comprehension are dissociable, and patients who have both word- and sentence-comprehension deficits typically have damage extending to subcortical white matter in addition to a temporoparietal lesion (Mesulam et al. 2015).

Furthermore, Calandri et al. (2014) have shown that bilinguals with PPA have greater microstructural integrity than monolinguals with PPA in the right uncinate fasciculus (UF). The UF has been proposed to play a role in lexical retrieval, semantic association, and aspects of naming that require connections from temporal to frontal areas which have been shown to undergo selective damage in the semantic variant of PPA. Calandri et al. (2014) suggest that the management of bilingual semantic knowledge could strengthen white matter pathways against degeneration, but suggest that further research is needed as this has just begun to be explored.

To summarize, PPA can provide crucial information about language organization over and above what we know from sudden-onset aphasia. Both the factors discussed above – gradual versus sudden loss, and different sites of neural damage – may provide a basis for differences in language impairment resulting from the two sources, such as language *loss* versus impaired language *access*. When language is suddenly impaired following a CVA, access to language is often due to disruption rather than loss. Therefore, full or partial recovery has the potential to occur, either spontaneously or with language treatment. In PPA, neural mechanisms can reorganize to at least partially compensate for language impairment. However, functions slowly degenerate, and the eventual decline will surpass compensation and result in the loss of language abilities, as seen in later stages of PPA. This language decline in PPA resulting from regional atrophy is less able to be recovered than language impairment resulting from a cerebrovascular lesion, and therefore intervention for PPA focuses on maintenance rather than restoration. It is important to mention that only one of the bilingual PPA studies investigated treatment effects on language (Meyer et al. 2015). The researchers found that the treatment did not restore function, but the treatment did slow decline, certainly for the treated language and possibly in the untreated language. Thus by studying language loss as a result of a degenerative disease, we can add to our knowledge of bilingual language organization on top of what we already know from studies of sudden-onset aphasia in bilinguals.

3. Support for Language Models

Based on the bilingual and multilingual PPA cases reviewed in Section 2, the patterns of language decline are consistent across the three variants of PPA. Notably, language decline occurs in all languages of bilinguals and multilinguals with PPA, relative to the specific atrophy of the variant of PPA, and performance on some tasks declines in parallel across languages even in the beginning stages. This provides some support for Green's (2003) convergence hypothesis, with processing for L2 and L1 converging on shared brain regions for both languages (Abutalebi 2008; Abutalebi and Green 2007; Green 2003). However, the convergence hypothesis is not fully supported, since in many cases, at least some language tasks decline differentially in the early and middle stages of the disease. Furthermore, proficiency and recency of use do not appear to have a substantial effect on language decline between the L1 and L2 in bilinguals and multilinguals with PPA, contrary to the convergence hypothesis. Recall that the only support for the convergence hypothesis came from contrasting two cases of multilinguals with PPA

(Kambanaros and Grohmann 2012; Mendez et al. 2004, participant 2). This will be an interesting direction of research in the future when more cases of multilinguals with PPA are investigated.

Regarding Ullman's (2015) declarative-procedural model, little support is found from the bilingual and multilingual cases of PPA that we have reviewed. Although differential decline is observed in some cases and some language tasks, with L1 better preserved than L2, the patterns of differential decline do not follow the hypotheses of the declarative-procedural model. A lower age of L2 acquisition did not consistently result in parallel decline, and differences between implicit and explicit learning contexts did not result in parallel vs. differential decline respectively. Furthermore, specific language tasks that are related more to declarative (explicit) learning in both the first language and second languages (e.g. lexical knowledge) relative to other tasks that are considered more procedural in the first language and more declarative in the second (e.g. syntactic knowledge) did not show parallel decline relative to differential decline as might be expected from the declarative-procedural model. To summarize, neither model is fully supported by the bilingual and multilingual PPA literature.

What is also notable about the PPA literature is the striking similarities between bilinguals with AD and those with PPA. When language declines in AD, L1 has been found to be better preserved than L2 with both languages declining in parallel at the end-stage of the disease. Language decline in PPA parallels this pattern, which is hypothesized to be due to the process of neural atrophy and its effect on language loss. The bilingual PPA literature, we would argue, can be particularly useful in understanding how language is organized in the brain, as AD patients typically have concomitant cognitive deficits at the late stages of the disease, which is when language decline is most noticeable. These cognitive deficits in AD may confound the picture of language decline, but in PPA – especially in the earlier stages of the decline – the picture may be clearer when the focus is specifically on language.

4. Conclusion

Studies on bilinguals with PPA can provide valuable information regarding language organization in the bilingual brain. Even with only 13 case studies covering a range of variants, languages, proficiency levels, ages of acquisition, manners of acquisition, and patterns of use, it is clear that the patterns of language decline fall into two main categories: parallel decline or L1 better spared than L2. Furthermore, as the PPA progresses, there is a trend towards parallel decline overall. Whether these two patterns will remain exclusive as more cases of bilinguals with PPA are studied and published, or whether there will be some cases of L2 being better spared than L1, remains to be seen.

For now, based on the published literature to date, we conclude that patterns of language decline in bilinguals with PPA are similar to those observed in bilinguals with AD; they do not support Ullman's (2015) hypothesis and only partially support Green's (2003) convergence hypothesis. More detailed cases of bilinguals with PPA are necessary to revise this, or other, models of bilingual language organization to fit better with the

PPA data. The degenerative nature of PPA, likely resulting in compensation for declining language abilities over the progressive decline, at least in the early stages, may explain why the two patterns of language decline converge to parallel decline over time. While the main factor driving the patterns of language decline is currently suggested to be order of acquisition, further research involving more cases of bilinguals with PPA, providing more details about language background and premorbid and postmorbid language abilities is essential. It is still unclear how other factors such as proficiency and/or dominance, age of acquisition, manner of acquisition, and patterns of use (such as recency of use) may interact with order of acquisition to determine the course of decline in any given bilingual with PPA.

In conclusion, longitudinal studies following the progression of the decline in both languages over time, including both behavioural and neuroimaging data, will be essential to understanding the progress of the degeneration, what compensation may occur, and how the deterioration affects languages in bilinguals with PPA. Further investigation of PPA in bilinguals should aid in our theoretical knowledge of language organization in the bilingual brain, particularly in areas that are still poorly understood, and should suggest avenues of approach for treatment to buffer against decline, enhance compensation, and maintain communication in order to uphold quality of life. The case studies that make up the current literature provide a first step towards understanding the complexity of language decline in bilinguals with PPA.

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