Journal of Critical Study of Communication and Disability

Shibata, K., Simpson, N., Reeder, J., Darby, R. R., & de Riesthal, M. (2024). Primary progressive aphasia and non-medical factors related to health outcomes: A retrospective study. *Journal of Critical Study of Communication and Disability*, 32–45. https://doi.org/10.48516/jcscd_2024vol2iss1.24

# Primary Progressive Aphasia and Non-Medical Factors Related to Health Outcomes: A Retrospective Study

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## Article Information

Keywords: Primary Progressive Aphasia, diversity, equity, inclusion, and justice, Care access, communication sciences and disorders, Neurodegenerative Disease

https://doi.org/10.48516/jcscd_2024vol2iss1.24

Submitted: July 21, 2023 EDT

Accepted: January 25, 2024 EDT

Published by Adelphi University Libraries

## Abstract

Primary progressive aphasia (PPA) is a neurodegenerative disease involving insidious onset language loss in the context of relatively spared cognitive function, where language is the initial and dominant factor impacting activities of daily living (Mesulam, 2001). Detection and identification of PPA requires a level of interdisciplinary expertise and resources that are not uniformly available across healthcare settings at the local and global levels (Bahia, 2007; Duran-Aniotz et al., 2021; Guimarães et al., 2013; Hogan et al., 2016; Knopman & Roberts, 2011; Onyike et al., 2020; Pijnenburg et al., 2004). This potential inequity raises concerns for increased risk of underdiagnosis, misdiagnosis, and/or delayed diagnosis of PPA, in turn impacting access to the required specialty care. The current study sought to characterize the PPA population at a Southeastern United States academic medical center, and to examine potential delays in PPA diagnosis and access to speech-language pathology services related to social determinants of health. We hypothesized that individuals from minoritized and otherwise disenfranchised groups would have longer duration of time between first reported symptom, and PPA diagnosis. Further, we hypothesized that these groups would be less likely to be referred to speech-language pathology services. Our results revealed a significant lack of sample diversity in terms of race, ethnicity, language practices, and queer identity. Within our overwhelmingly White, non-queer-identifying sample, factors related to rurality and economic status did not predict time to diagnosis or referral to speech pathology. A discussion of PPA and related disorders as they relate to social determinants of health follows.

Equitable access to quality healthcare is a fundamental human right. Health equity can be appreciated when outcomes are consistently resistant to the influence of non-medical, socially derived factors such as race, ethnicity, languaging practices, gender expression and queerness, economics, education, and geographic location. Upon the backdrop of a for-profit healthcare system in a country founded upon patriarchy and White supremacy, the existence of significant health disparities along the lines of these socially derived factors in the United States (US) is unsurprising (e.g., Kendi, 2019; Zinn, 1980). Dementia spectrum disorders are among the top 10 causes of death in the US regardless of race, ethnicity, or sex assigned at birth (Centers For Disease Control, n.d.). Nonetheless, health disparities in dementia endure along aforementioned social lines, especially race and ethnicity (e.g., Cooper et al., 2010; Mehta & Yeo, 2016). Importantly, the US population is expected to increase significantly in the coming decades, and at remarkably higher rates within minoritized populations. We anticipate that more people will be living with dementia, and that more of them will be non-White (Matthews et al., 2018).

One variant of dementia, known as primary progressive aphasia, is the focus of the current study. Primary progressive aphasia (PPA) is a neurodegenerative disease involving insidious onset language loss in the context of relatively spared cognitive function, where language is the initial and dominant factor impacting activities of daily living (Mesulam, 2001). Detection and identification of PPA requires a level of interdisciplinary expertise and access to resources not uniformly available across healthcare settings on both local and global levels (Bahia, 2007; Duran-Aniotz et al., 2021; Guimarães et al., 2013; Hogan et al., 2016; Knopman & Roberts, 2011; Onyike et al., 2020; Pijnenburg et al., 2004). This potential inequity raises concerns for increased risk of underdiagnosis, misdiagnosis, and/or delayed diagnosis of PPA, impacting access to the required specialty care. In the following sections, we contextualize the interplay between challenges in the detection and diagnosis of PPA and non-medical socially derived factors related to health outcomes (henceforth referred to as non-medical factors). We will first outline the diagnostic complexities in

**Table 1. Clinical Diagnostic Criteria of the Primary Progressive Aphasias**

| Feature Type | nfvPPA | svPPA | lvPPA |
|---|---|---|---|
| Core Features | At least one core feature, and two additional features must be present | Both core features, and at least three additional features must be present | Both core features, and at least three additional features must be present |
| | Agrammatism in language production | Impaired confrontation naming | Impaired single-word retrieval in spontaneous speech and naming |
| | Effortful, halting speech with inconsistent speech sound errors and distortions (apraxia of speech) | Impaired single-word comprehension | Impaired repetition of sentences and phrases |
| Additional Features | Impaired comprehension of syntactically complex sentences | Impaired object knowledge, particularly for low-frequency or low-familiarity items | Speech (phonologic) errors in spontaneous speech and naming |
| | Spared single-word comprehension | Surface dyslexia or dysgraphia | Spared single-word comprehension and object knowledge |
| | Spared object knowledge | Spared repetition | Spared motor speech |
| | | | Absence of frank agrammatism |

*Note.* nfvPPA = Nonfluent/agrammatic primary progressive aphasia; svPPA = Semantic variant primary progressive aphasia; lvPPA = Logopenic variant primary progressive aphasia (Gorno-Tempini et al., 2011)

PPA, followed by a discussion of each non-medical factor as it relates to PPA and related disorders.

## Diagnostic Complexities in Primary Progressive Aphasia

Primary progressive aphasia is most frequently diagnosed and managed by a fellowship trained cognitive behavioral neurologist through a specialty clinic at an academic medical center. At minimum, a PPA diagnosis requires a medical neurological workup including clinical and radiographic assessment, but may also include input from a neuropsychologist or speech-language pathologist. Gorno-Tempini et al. (2011) produced the seminal international consensus paper, which established behavioral, neuroimaging, and pathophysiological diagnostic profiles for three variants of PPA: nonfluent/agrammatic variant (nfvPPA), semantic variant (svPPA), and logopenic variant (lvPPA). Table 1 illustrates the current consensus on diagnostic criteria. It should be noted that many practitioners recognize a distinct neurodegenerative speech process that involves impaired motor planning for speech production with spared language skills, termed primary progressive apraxia of speech (Duffy, 2006). Alternatively, other practitioners contend that this presentation is a sub-variant of nfvPPA.

A clinical diagnosis of PPA ideally includes comprehensive, interdisciplinary assessment of behavioral, cognitive, language, and motor domains to ensure diagnostic accuracy. Expert consensus recommends thorough language assessment by providers with expertise in evaluating and treating individuals with this diagnosis (Gorno-Tempini et al., 2011). This is of particular importance in the initial stages of the disease when the symptoms are mild and more difficult to detect. Diagnostic certainty is supported by imaging techniques such as magnetic resonance imaging (MRI), positron emission tomography (PET), or single-photon emission computed tomography (SPECT). Importantly, atrophy is insidious and not typically noted by radiologists given the need to corroborate evidence of atrophy with clinical presentation. Thus a neurologist experienced with PPA must review and interpret imaging results in the context of clinical presentation. Testing for a neurodegenerative disease-associated proteinopathy or pathogenic mutation can also increase diagnostic certainty. Taken together, these diagnostic requirements tend to restrict proper PPA diagnosis and management to specialty clinics, usually within academic medical centers.

## Race and Ethnicity

Due to a variety of issues, prevalence of PPA is difficult to surmise at the population level. The limited evidence available suggests a prevalence around 3-4/100,000 (Coyle-Gilchrist et al., 2016; Knopman & Roberts, 2011; Mesulam et al., 2021). To our knowledge no information is available on the prevalence of PPA within groups of the U.S. population. Rates of general dementia prevalence may give some insight into potential disparities along the lines of race and ethnicity. Dementia prevalence has been shown to be moderated by immigrant status, except for Black Americans for whom prevalence and lifetime dementia burden is the highest, especially

among Black men (Farina et al., 2020; M. A. Garcia et al., 2019; Moon et al., 2019). Garcia et al., (2019) also found that Latinx/a/o Americans born domestically bear an increased lifetime dementia burden regardless of gender. Limited statistics exist for people indigenous to lands occupied by the US, but available evidence suggests that dementia burden is higher for indigenous populations residing in occupied lands within and outside US territories (Mayeda et al., 2016; Warren et al., 2015). Information about Asian Americans and Pacific Islanders is also unclear, with some studies reporting lower prevalence and others suggesting higher prevalence of dementia and mild-cognitive impairment (Hayes-Larson et al., 2022; Mehta & Yeo, 2016). The term "Asian American", originally coined by Bay Area Asians from diverse ethnic groups to capture the reductive generalizations made against them by White Americans, may contribute to this confusion (Umemoto, 1989). In the US, Asian American refers to a broad group of individuals without differentiation between ethnic groups, whose sociopolitical histories differ significantly from each other. Evidence from ethnically diverse Asian countries (e.g., Singapore) suggest dementia and neurodegenerative disease prevalence differs among minoritized and dominant Asian groups (Ng et al., 2007; Sahadevan et al., 2008).

## Languaging Practices

According to the latest figures, nearly 20% of the U.S. population speaks a language other than English at home (US Census Bureau, 2022b), strikingly less than the global population's bilingualism rate of more than 40% (Ansaldo et al., 2008). Nonetheless, careful consideration of the potential impact of languaging practices on the diagnostic process for PPA is warranted. Individuals with PPA who use two or more languages in their daily lives (i.e., translanguagers [O. Garcia, 2011]) exhibit heterogeneous profiles of decline across languages, but typically the first acquired language remains or becomes the strongest (Malcolm et al., 2019). Similarly, Mendez et al., (2020) found that translanguagers with Alzheimer's disease (AD) (the underlying etiology of many PPAs) demonstrated delayed symptom onset by about 4 years, and that a large proportion of individuals had reverted to their first acquired language after disease onset. As such, there is potential for diagnostic delays or inaccuracies if there are no available experts that can communicate with and evaluate an individual across their languaging domains.

Of course, not all individuals with PPA in the US include English in their languaging practice. A recent systematic review of language barriers in healthcare settings demonstrated that language barriers between patient and provider contributed to overall decreased satisfaction, decreased quality of care, and increased safety risks (Shamsi et al., 2020). The same review suggested increased safety and satisfaction with online translation platforms such as Google Translate and MediBabble, however these platforms are not conducive to implementation of communication strategies often required in the case of PPA and other neurogenic communication changes (e.g., individuals with non-fluent/agrammatic PPA require modification of sentences to simplified grammar structures). Similarly, data from other neurology subspecialties, such as stroke and movement disorders, suggest higher levels of communication disorder severity at discharge and lower levels of knowledge and care seeking by individuals who do not speak the language in which care is provided (Pan et al., 2014; Shah et al., 2015). Perhaps most revealing is the recent work by Alexandra Miner (2022) which elucidates barriers to neurological care for groups who do not use English. Through retrospective data analysis, Miner found lowest rates of outpatient follow up in traumatic brain injury, dementia, and pain syndromes, and high rates of psychiatric and neurological comorbidities across etiology. In qualitative interviews with patients, themes of hesitancy to ask questions, socioeconomic barriers that prevent individuals from prioritizing healthcare, confusion about diagnoses and medication, and overall low expectations for care and outcomes were conspicuous (Miner, 2022).

## Sex, Gender, and Queer Identities

Although cisgender men and cisgender women appear to be equally affected by PPA, there are no reports of the impact of PPA on individuals for whom the gender binary excludes, and the unique experiences of queer-identifying individuals living with PPA is absent from the literature. In terms of general dementia, many have identified that queer folx are routinely underserved (e.g., Cousins et al., 2020; Di Lorito et al., 2021; Harper, 2019). Others, using queer and crip theory, have argued that the way person-centered approaches in dementia care are typically carried out can be harmful, reproduce heteronormativity, and subject individuals with dementia to regendering (Foth & Leibing, 2022; King, 2021). Guo et al., (2022) examined dementia risk in transgender individuals by comparing 1,784 transgender adults with age and race matched controls (each transgender individual was matched with 10 cisgender women and 10 cisgender men). They found that transgender individuals were more likely to develop dementia and were more likely to have positive risk factors for dementia development in all but two measured categories. That is, when comparing transgender individuals and cisgender controls, there was a statistically significant difference in prevalence of smoking and alcohol abuse history, depression, hearing loss, sleep disorders, and factors related to cardiovascular health. Moreover, cisgender women are disproportionately affected by lifetime dementia burden (i.e., the cumulative effect of disease prevalence, incidence, severity, financial cost, and impact on daily life), and most caregivers of individuals with dementia (including PPA) are women (Alzheimer's Association, 2019). Mayeda (2019), points out that the conflation between biological sex assigned at birth (i.e., sex) and gender identity, (i.e., gender) a social construct, leads to difficulty in exploring mechanisms contributing to the disproportionate impact of dementia on cisgender women and others that the gender binary excludes. Conflating these terms prevents eval-

uation of risk factors that could be specific to sex assigned at birth, effects of the social construct of gender, or other treatments that individuals may undergo during their lifetime to affirm their gender identity (e.g., hormone therapy).

## Geographic Location

The constellation of information required to diagnose PPA calls for a collaborative, interdisciplinary team of professionals with special expertise in assessment, treatment, and/or management of the disease and its sequelae. Acquisition of brain imaging, histopathological findings, and/or genetic information may also be warranted. These prerequisites are most commonly satisfied by large, well-funded academic medical clinics and research centers, restricting PPA care to such institutions. Most clinical-research centers concerned with PPA and its related disorders are concentrated within major metropolitan areas in North America and Europe (Onyike et al., 2020). Furthermore, it has been demonstrated that individuals residing in rural areas are at higher risk of developing dementia due to a variety of environmental and situational factors such as limited access to care providers and toxic environmental exposure (Aranda et al., 2021). Individuals outside the catchment area of specialty centers may be at a disproportionate risk of underdiagnosis, misdiagnosis, and/or delayed diagnosis of PPA. Another contributing factor to diagnostic issues is the readiness to independently seek specialty care without referral. Individuals in rural areas are less likely to self-refer to specialists, and more likely to live in areas where PPA care does not exist (Bellinger et al., 2010). Physically reaching neurological services (spatial access) has been identified as a major threat to care outcomes in some of the US' most spatially vulnerable populations (Buchalter et al., 2023).

## Economic Factors

Economic diversity in the US is extremely variable, with an official poverty rate of 11.4% juxtaposed with the top 1% of earners in the US who earn a minimum income of more than $420,000 annually and an average income of more than 1.3 million dollars annually (*Interactive: The Unequal States of America*, n.d.; US Census Bureau, 2022a). Several studies from Western nations, all outside of the US, have explored the relationship between economic factors, such as household income and education, with dementia severity at time of diagnosis. Qian and colleagues (2014) conducted a retrospective study with data from their memory disorders clinic in Toronto and found that patients with lower socioeconomic status (proxied by educational attainment and annual income) were more likely to receive a diagnosis of Alzheimer's versus mild cognitive impairment (MCI), were older at time of diagnosis than individuals diagnosed with MCI, and had lower use of drugs that enhance cognition in dementia. In Denmark, a country with socialized healthcare and presumed access equity relative to the US, Petersen and colleagues (2021) found that a higher household income was associated with more mild symptoms at the time of dementia diagnosis. These studies suggest that patients with more economic resources are getting into clinic sooner in the disease process, and accessing resources and care at more mild stages of the disease, when certain drugs and interventions are most effective. Coupling the economic diversity in the US with the landscape of a for-profit healthcare model, one may ponder its impact on access to dementia diagnosis and care. While a variety of studies have been conducted on ongoing dementia care access, dementia diagnosis as it relates to economic factors is largely unexplored in the literature. Unsurprisingly, these queries are unaddressed within the PPA literature.

Although insufficient information exists related to dementia diagnosis and economic factors in the US, it is well documented that dementia care costs in the US are high on healthcare systems, high on payors, and highest on families (e.g., Aranda et al., 2021; Jutkowitz et al., 2017; Kelley et al., 2015). Of paramount significance for individuals with PPA is the fact that typical onset is before age 65, before most people have retired, and before they are eligible to receive federal medical benefits available to all US residents who are citizens or documented residents (known as Medicare). Kelley et al. (2015) point out that even under Medicare, home care services, adaptive equipment, and residential care are generally not covered, down-streaming costs to families to the point that by the time of death, more than half of all patients with dementia qualify for the US federal healthcare plan reserved for very low-income families (known as Medicaid). That is, more than half of all patients with dementia will be considered "very low-income" at the time of their death. This proportion increases to three-fourths for individuals from minoritized communities, and ultimately places informal caregiving responsibilities on families, leaving families from minoritized groups disproportionately impacted ( Alzheimer's Association, 2018; Schulz & Beach, 1999).

## The Current Study

The complexities of these diagnostic criteria held constant, the influences of non-medical factors on the diagnosis of PPA remain opaque. This is of particular importance in the context of the United States, where population diversity is ever increasing. Economic resources and rurality also vary widely across the US and have a longstanding relationship with access to care (Bellinger et al., 2010; Franks & Clancy, 1997). The heterogeneous manifestations of PPA symptoms superimposed upon a landscape of convoluted social characteristics and conditions leave PPA uniquely vulnerable to diagnostic delays, underdiagnosis, and misdiagnosis.

The current study sought to explore the potential impact of socially derived non-medical factors related to health outcomes on diagnosis timeliness and receiving a referral to speech therapy for individuals with PPA within a Southeastern US academic medical center. As dis-

cussed above, non-medical factors, such as race, ethnicity, languaging practices, queer identities, rurality, and economic resources intersect with health outcomes, and may be especially important in the context of medically complex or rare disorders such as PPA. The diagnostic complexity of PPA provides a uniquely threatening foreground for diagnostic timeliness and accuracy, leaving patients vulnerable to the impact of aforementioned non-medical factors. We hypothesized that minoritized group membership, further distance from clinic, and lower socioeconomic status would predict a longer time between symptom onset and PPA diagnosis, and that these individuals would be less likely to be referred to speech therapy services. To test these hypotheses, we asked, within a Southeastern US academic medical center:

1. What non-medical factors related to health outcomes characterize individuals diagnosed with PPA?
2. Do specific non-medical factors predict a longer time to PPA diagnosis?
3. Do specific non-medical factors predict referral patterns to speech-language pathology?

## Method

### Setting

This study was conducted using clinical data derived from the electronic medical record at Vanderbilt University Medical Center (VUMC). VUMC is in Nashville, Tennessee which is located in Southern United States. It provides services to individuals in all 95 counties of the state, and specialty care to a large proportion of the region, drawing from a 65,000 square mile catchment area that spans Northern Alabama, all of Tennessee, and Southern Kentucky (VUMC Office of Government and Community Affairs, n.d.). VUMC is the only institution offering specialty care for frontotemporal lobar degeneration (FTLD) and PPA in the state, and borders several states that have no options for FTLD or PPA specialty care (The Association for Frontotemporal Lobar Degeneration, 2022). As such, individuals with PPA symptoms are referred from across the region for assessment and management of PPA and other FTLD spectrum disorders. This allowed for retrospective analysis of a large sample size of individuals diagnosed with PPA relative to population prevalence. Before proceeding with data collection, we estimated the distribution of individuals served at VUMC by race and ethnicity to ensure that the demographic distribution of the hospital population reflected the demographics of Tennessee. To do so, we used the synthetic derivative, which is a system at VUMC that maintains a de-identified copy of the medical record (Danciu et al., 2014; Roden et al., 2008). It offers the ability to generate approximate counts of variables extracted from the electronic medical record, including demographic characteristics and diagnosis code. We were unable to use the synthetic derivative to generate a priori estimates of the ethnoracial distribution within the PPA cohort because PPA does not have a unique diagnostic code.

### Data Acquisition

In order to address our research questions, medical records of individuals with a diagnosis of PPA and PPAOS who were seen between November 2017 and April 1, 2022, were identified in the Vanderbilt University Medical Center electronic medical record system. Internal Review Board approval was obtained through Vanderbilt University. A diagnosis of PPA was determined by fellowship-trained cognitive behavioral neurologists, often in collaboration with related professionals such as neuropsychologists or speech-language pathologists. Individuals were identified through searching the medical record by ICD-10 code combinations and problem list combinations. To identify patients in the system who carried a PPA diagnosis, the medical record automatically pulled all patients with a diagnosis of both a communication problem and frontotemporal lobar degeneration, or a communication problem and Alzheimer's disease. Additionally, all patients who had a diagnosis of a communication problem and had been seen by a cognitive behavioral neurologist were reviewed for selection. Communication problem was defined as any one, or combination of the following diagnoses: primary progressive aphasia, aphasia, primary progressive apraxia of speech, apraxia of speech, speech problem, and/or communication impairment. Each medical record was then manually reviewed by two independent coders (authors KS and NS) to ensure that the diagnosis was indeed primary progressive aphasia, and not another type of aphasia in the setting of dementia. A final list of records to include and exclude were reviewed by consensus meeting between the two coders. If there was not agreement on a particular record, it was independently reviewed by a third coder (author MdR) for discrepancy resolution.

Variables related to medical diagnosis of PPA and non-medical factors related to health outcomes were extracted via electronic medical record-generated reports and manual review of the medical record between April 1, 2022, and July 31, 2022. Two independent coders (authors KS and NS) extracted these variables through manual chart review. Discrepancies were resolved through collaborative consensus between the two coders, with a third independent coder (author MdR) in place to resolve final discrepancies if needed. Many target variables included in the study are not reliably or systematically collected across the medical center, requiring manual extraction from the electronic medical record. PPA evaluation requires neurologists to take a detailed, thorough history from a patient and their family, so neurologist's notes typically contain rich information, both medical and non-medical. Nonetheless, we anticipated and planned for missing data by running a prospective power analysis using SPSS Statistical Software by IMB (Version 29.0). We estimated the number of observations needed to run a multiple regression model based on a varying number of predictors, and concluded that despite the

inevitability of missing data, we were likely to derive enough power to proceed with a regression model using at least a subset of non-medical factors as predictors.

Variables related to the medical diagnosis of PPA that were manually extracted from the record, include PPA diagnosis subtype, date of clinical PPA diagnosis, date of imaging or pathology supported diagnosis, date of first reported symptom by both patient and family member/care partner, and consideration of referral to speech-language pathology services. Non-medical variables related to health outcomes included sex assigned at birth, race, ethnicity, preferred language, all of which were generated by system reports. Of note, race identity options offered to patients were limited to Black, White, Asian/Pacific Islander, people indigenous to lands occupied by the US, and mix race. Ethnic categories offered followed US Census Bureau convention which is limited to a binary choice of Latinx/a/o, or non-Latinx/a/o White due to a 1976 law passed by the US Congress that selectively required tracking of individuals of Spanish-speaking background (Lopez et al., 2023). Additionally, factors associated with socioeconomic status were manually extracted and included median income by zip code, years of education, highest level of education, occupation type and occupational status. Gender identity and sexual orientation were manually extracted from the medical record, where available. Distance to clinic was used as a proxy for rurality and was derived by calculating the distance between patient's zip code and Vanderbilt University's main medical campus on Google Maps.

## Results

### Research Question One

To better understand the non-medical factors contributing to the backgrounds of individuals with PPA who are diagnosed and cared for at this Southeastern US academic medical center, our first question aimed to generate detailed descriptive statistics. We identified 143 patients in the medical record who were diagnosed with PPA between November 2017 and April 1, 2022. Descriptive statistics were derived using R Statistical Software (R 4.3.1; R Core Team, 2021), and detailed results are displayed in Table 2. Overall, individuals diagnosed with PPA were overwhelmingly White and monolingual. Other variables that served as socioeconomic proxies (medium income by zip code, education, distance from clinic, and population by zip code) were evenly distributed. Notably, the sample demonstrates an underrepresentation of several minoritized groups compared with broader Tennessee statistics (US Census Bureau, n.d.). Underrepresented minoritized groups included those who identify as queer and gender diverse, Black, Latinx/a/o, and those with languaging practices that included non-English languages. This prevented us from including these non-medical variables in the statistical models for our second and third research questions due to insufficient power.

### Research Question 2

Our second question sought to evaluate whether non-medical factors related to health outcomes predicted unique variability in the time between symptom onset and receiving a diagnosis of PPA. A multiple linear regression was carried out using R Statistical Software (R 4.3.1; R Core Team, 2021). Sufficiently powered predictor variables included were level of education (by highest attained degree), median income by zip code, population by zip code, and how far from the medical center individuals resided. Our outcome variable was the time between first reported symptom, and the date of a clinical PPA diagnosis in months (M= 42.89, SD= 31.70). Although the outcome variable's distribution was non-normal, linear regression models are generally robust to violations of normality (Cohen et al., 2014). To confirm this, we visually analyzed diagnostic plots, all of which were in the acceptable range (i.e., residuals were normally distributed). Ultimately the model was non-significant ($F_{9,99} = 0.487$, $p = 0.880$). In other words all predictors combined did not predict unique variability in the time between symptom onset and diagnosis.

### Research Question 3

Our third question queried whether non-medical factors predicted variability in speech language therapy referrals. Again, we ran a multiple linear regression using R Statistical Software using the same predictor variables. The model was non-significant ($F_{8102} = 0.608$, $p = 0.769$), indicating that all predictors combined did not predict referrals to speech language therapy after a diagnosis of PPA.

## Discussion

To our knowledge, no data exists characterizing the social demographic makeup of individuals diagnosed with PPA, and the influence of these factors on diagnosis and treatment remains unexplored. The current study aimed to better understand the characteristics of patients with PPA who received care at one of the few specialty sites for PPA in the Southeastern US through the lens of non-medical socially derived factors related to health outcomes. Further, we sought to determine if non-medical factors predicted variability in the timeliness of an individual receiving a PPA diagnosis. Results were limited by an overall lack of representation of minoritized groups in terms of race, ethnicity, languaging practices, and queer identities. Our sample did contain a relatively diverse array of educational levels and median incomes by zip code, which serve as proxies for socioeconomic status. Thus, our regression models' predictors included factors related to economic status and rurality, and both models were ultimately non-significant. Implications of these findings are discussed in the following section.

Regarding our first research question, results of descriptive statistics in terms of race and ethnicity are in conflict with the demographics of local and state popula-

**Table 2. Descriptive Statistics**

| Primary Progressive Aphasia Subtype (n=143) | | | | |
|---|---|---|---|---|
| Category | n | Percent of sample | | |
| lvPPA | 87 | 61% | | |
| svPPA | 25 | 17% | | |
| nfvPPA | 16 | 11% | | |
| PPAOS | 4 | 3% | | |
| PPA NOS | 6 | 4% | | |
| notPPA | 5 | 4% | | |
| Race (n=143) | | | | |
| Category | n | Percent of total sample | Percent of reported sample | Percent of TN Population |
| Black | 4 | 2.7%** | 3.4%** | 16.7% |
| White | 112 | 78% | 96.5%** | 78.3% |
| Asian/Pacific Islander | 0 | 0% | 0% | 2.2% |
| People indigenous to lands occupied by the United States | 0 | 0% | 0% | 0.5% |
| Mix race | 0 | 0% | 0% | 2.2% |
| Unknown | 27 | 19% | NA | NA |
| Ethnicity (n=143) | | | | |
| Category | n | Percent of total sample | Percent of reported sample | Percent of TN Population |
| Latinx/a/o | 0 | 0%** | 0%** | 6.4% |
| Non-Latinx/a/o White | 93 | 65%** | 100%** | 72.9% |
| Unknown | 50 | 35% | NA | NA |
| Sex Assigned at Birth and Queer Identities | | | | |
| Category | n | Percent | | |
| Female | 81 | 57% | | |
| Male | 62 | 43% | | |
| Intersex | 0 | 0% | | |
| Queer Identities | 0 | NA | | |
| Highest Attained Degree (n=113) | | | | |
| Category | n | Percent | Percent of TN Population over 25 years of age | |
| Less than High School | 3 | 3% | 9.6% | |
| High School Diploma or GED | 27 | 24% | 31.3% | |
| Two-Year Degree or Training | 28 | 25% | 27.9% | |
| Bachelor's degree | 25 | 22% | 19.7% | |
| Master's Degree | 18 | 16% | | |
| Doctorate or Higher | 12 | 10% | | |
| Master's Degree or higher | 30 | 27%** | 11.4% | |
| Occupational Status (n=126) | | | | |
| Category | n | Percent | | |
| Employed | 9 | 7% | | |
| Retired | 67 | 53% | | |

| | | | |
|---|---|---|---|
| Unable to work (due to PPA) | 44 | 35% | |
| Not in the workforce (e.g., "homemaker") | 6 | 5% | |
| Age | | | |
| Minimum | 50 | | |
| First Quartile | 67 | Mean | 71.89 |
| Median | 72 | | |
| Third Quartile | 78 | | |
| Maximum | 87 | | |
| Distance to Clinic (mi) | | | |
| Minimum | 1.2 | | |
| First Quartile | 15.25 | Mean | 79.32 |
| Median | 36.5 | | |
| Third Quartile | 117.5 | | |
| Maximum | 827.0 | | |
| Population | | | |
| Minimum | 794 | | |
| First Quartile | 11,675 | Mean | 27,401 |
| Median | 22,000 | | |
| Third Quartile | 40,295 | | |
| Maximum | 64,860 | | |
| Median Annual Income | | | |
| Minimum | $38,994 | | |
| First Quartile | $49,824 | Mean | $61,789 |
| Median | $62,515 | | |
| Third Quartile | $69,298 | | |
| Maximum | $131,715 | | |

*Note*. lvPPA = Logopenic variant primary progressive aphasia; svPPA = Semantic variant primary progressive aphasia; nfvPPA = Non-fluent/agrammatic variant primary progressive aphasia; PPAOS = Primary progressive apraxia of speech; PPNOS = Primary progressive aphasia not otherwise specified; NotPPA = Initially thought to be primary progressive aphasia but diagnosis was later overturned; TN = Tennessee; PPA = Primary progressive aphasia; GED = General educational development test

*Percentages cannot be derived from the umbrella category queer identity, as it refers so anyone who does not identify as cisgender or heterosexual, thus individuals may identify with several groups, and it is distinct from sex assigned at birth.

** Statistically significant difference when compared to the greater Tennessee population, as indicated by significant p-value of greater than 0.05

tions. According to the latest census data, the Nashville population is 27.2% Black and 60.5% White and the Tennessee population is 16.7% Black and 78.3% White (US Census Bureau, n.d.). However, as outlined in Table 2, of the individuals diagnosed with PPA in our sample, only 2.8% (4 individuals) of patients were Black, and 78% (112 individuals) were White, with 18.88% unreported. No patients were identified in the record as Asian/Pacific Islander, people indigenous to lands occupied by the US, or Latinx/a/o. To further contextualize, we examined the demographics of individuals diagnosed with diseases underlying PPA (Alzheimer's disease and frontotemporal lobar degeneration) at Vanderbilt University Medical Center in a post-hoc analysis using the synthetic derivative. We found that the racial distribution within the group of individuals what carried diagnoses that underly PPA is slightly incongruent with the broader population demographics for Tennessee (which the latest census reports as 13% Black, 81.4% White), the findings are not nearly as disproportionate as the PPA population. As outlined in the introduction, these results are also in conflict with broader dementia and Alzheimer's research which suggest rates of dementia are higher in minoritized groups (Alzheimer's Association, 2019; Aranda et al., 2021; Cousins et al., 2020; Di Lorito et al., 2021; Farina et al., 2020; M. A. Garcia et al., 2019; Guo et al., 2022; Hayes-Larson et al., 2022; Matthews et al., 2018; Mayeda et al., 2016; Moon et al., 2019; Ng et al., 2007; Sentell et al., 2015). Overall, our findings demonstrate a concerning lack of representation of individuals from minoritized communities and suggest that further research is needed to characterize potential health inequities within the PPA population. While one may be tempted to query whether PPA is a disease that only impacts those of European decent, various genetic mutations have been identified and documented in non-white participants, and cases of PPA and related diseases have been documented around the world (e.g., Kawakatsu et al., 2021;

Kim et al., 2022; Majounie et al., 2012; Onyike et al., 2020; Xu et al., 2023). In fact, 30 years prior to emerging descriptions of svPPA in Western countries, neuropsychiatrist Dr. Tsunero Imura described the symptomatology of svPPA within Japanese cohorts, which he termed Gogi aphasia (literally, "word-meaning" aphasia). This leaves us to question: Where are all the BIPOC folx with PPA, and why are we missing them?

Linguistic diversity in our sample was also lacking. All patients had a documented preferred language of English, and 99% (141 individuals) of our sample reported using only English in the home. While languaging practices in Tennessee are less diverse than the population average in the US, these data remain incongruent to local and state data. According to the most recent release of American Community Survey data, 88.6% of individuals in Davidson County and 96.2% of individuals in Tennessee report using only English in the home (US Census Bureau, 2022b).

Additionally, our sample did not include anyone who openly shared a queer identity. The estimated percentage of individuals who identify as queer in Nashville is small to begin with (3.7%) and given the hostile anti-queer sociopolitical climate in the state of Tennessee, it is unclear how many individuals would openly identify as queer in their healthcare setting (Conron et al., 2020; *Human Right Campaign Tennessee Scorecard*, n.d.). Additionally, Vanderbilt University Medical Center did not have a dedicated space to report queer identities in the electronic medical record until 2020, which occurred during the period from which the sample was derived from (Vanderbilt University, 2023).

Our second and third research questions must be discussed in the context of the findings from research question one. We were unable to include non-medical factors related to ethnoracial, languaging, and queer identities, which have all been associated with differential dementia prevalence and outcomes (as outlined in the introduction). Our sample did, however, present an opportunity to evaluate the impact of proxies related to economic and geographic location factors (i.e., highest attained degree, median income by zip code, distance to clinic, and population size by zip code) in a homogeneous sample of individuals diagnosed with PPA that was overwhelmingly White, non-Latinx/a/o, and monolingual. For this group of individuals, proxies for economic and geographic location factors did not predict time to diagnosis, nor did they predict referral to speech therapy. This suggests that although geographic location and economic resources are important factors in dementia diagnosis and management, they do not appear to have the same impact on individuals with PPA who are White, non-Latinx/a/o, and monolingual. Would these finding hold true for individuals with PPA who *are* members of minoritized groups?

Fundamentally, the outcome variables in this study (time to diagnosis and referral to speech) are related to care access: Who can get a timely diagnosis, and who can get therapy for it? Situating these queries within the discussion of our sample's homogeneity, this study suggests that minoritized groups may have less access to PPA diagnosis and care. Studies of broader neurology specialty care access and utilization suggests that minoritized ethnoracial groups are diagnosed later in the disease process and less likely to receive antidementia medication, clinical trial opportunities, 24-hour-care services, and outpatient services (versus emergency and acute services) (Aranda et al., 2021; Cooper et al., 2010; Saadi et al., 2017). The same literature also suggests that the reduced access experienced by minoritized ethnoracial groups (even when controlling for economic factors) are also experienced by individuals who have fewer years of formal education and/or live in rural communities. Notably, however, our sample included a broad distribution of individuals across education levels, income levels (by zip code), and ruralness, but none of these factors predicted time to diagnosis or referral to speech therapy in this otherwise homogeneous PPA cohort. Ultimately, reduced access to PPA diagnosis and specialty care also restricts research opportunities, and inherently produces bias in the body of knowledge that informs our clinical decisions. Only individuals with access are potentially eligible to participate in contributing to this body of knowledge. In other words, the clinicopathological substratum upon which we base our "empirical" conclusions is made up exclusively by those who were able to overcome the barriers to subspecialty access. In turn, we generalize the outcomes of these studies to all individuals with PPA, often without evaluating the influence of non-medical factors unique to the individual.

Taken together, the lack of diversity in the current study sample and the non-significant impact of economic and geographic location factors on time to diagnosis and referral to speech for our homogenous sample raise disconcerting questions about equity in access to care and prompts ideas for important future directions. Most importantly, we need prospective studies carried out by diverse research teams in culturally and linguistically relevant and meaningful ways that focus on assessing and increasing PPA care access to all groups. Future studies should include non-medical factors in their reported demographic information in order to understand how non-medical factors intersect with PPA prevalence, care and outcomes. Currently, our research team is replicating this study at a West Coast academic medical center with a larger and more diverse cohort that may permit answering the lingering question of potential disparities in time to diagnosis and referral to treatment in minoritized ethnoracial groups. Additionally, we are reviewing PPA treatment literature to examine reporting practices and representation of minoritized groups within research cohorts to better understand the population we base our behavioral treatments upon.

## Limitations

A major limitation of the current study is the retrospective design. Variables were collected in a clinical context without consideration of scientific rigor, and with inevitable variability between practitioners and staff

members. Many of the variables collected were manually extracted from the medical record, usually from cognitive behavioral neurology reports that provided narrative information about individuals' social and non-medical environments. Thus, variables were not systematically collected and susceptible to clinician and staff variability and error. Additionally, we used proxies to capture certain factors, such as median income by zip code to estimate socioeconomic status, which are often crude and limited measures. Further, because this was a retrospective study without a consent process or protocol, it is reasonable to assume that some individuals chose not to disclose certain non-medical factors, especially identities that have historically and/or are currently suffering persecution within the Southeastern region of the US (e.g., queer folx, and non-White individuals, especially Black Americans and people indigenous to lands occupied by the US).

## Conclusion

Primary progressive aphasia is a complex, rare, heterogeneous disease with unknown prevalence within minoritized groups. The current study identified that individuals from minoritized groups are underrepresented in the PPA population within one large academic medical center in the Southeastern United States. Further, within our homogenous sample, we found that proxies for socioeconomic status and geographic location did not predict variability in the time between an individual's first reported symptom and receiving a diagnosis of PPA. As we continue to investigate and characterize the disease process of PPA from a biomedical perspective, we cannot ignore the socially derived non-medical factors related to health outcomes. Understanding the way these factors converge to impact PPA prevalence, care, and outcomes is critical to rendering intentional, equitable service provision.

## Positionality Statements

### *Kiiya Shibata, M.S., CCC-SLP (she/they)*

Kiiya Shibata is a half White Irish American, half Japanese Nisei (i.e., father was an immigrant from Japan), bilingual, non-disabled, queer woman/person, born and raised in Northern San Diego, California. Her identity and experiences as a mix-race, mix-lingual, mix-sexuality individual has driven her clinical and research interests toward intersectional spaces.

### *Nickolas Simpson, B.A. (he/him)*

Nickolas Simpson is a white, cisgender, man who was born and raised in multiple regions across the United States. As the son of two medical workers with consistent access to healthcare, his knowledge of health disparities comes primarily from his professional work, rather than personal experience.

### *Jerica Reeder, B.S. (she/her)*

Jerica Reeder (She/her) is an American born, white, cisgender, heterosexual woman who was raised in the depths of rural Tennessee. She has witnessed the lack of education and resources regarding one's health care journey, especially therapeutic outlets such as speech-language, mental health, and community support. This drives her ambition to advocate for advancing communal access to healthcare opportunities.

### *R. Ryan Darby, MD (he/him)*

Ryan Darby is an American born, English-speaking white male neurologist. As a physician, he recognizes that he is in a position of privilege with high healthcare literacy and easy access to healthcare resources, which may significantly contrast with the more limited healthcare literacy and access available to marginalized persons that he may evaluate and treat.

### *Michael de Riesthal, PhD, CCC-SLP (he/him)*

In contributing to the analysis and interpretation of this manuscript, he acknowledges his standpoint as a non-disabled, middle-age White cisgender male from Long Island, NY, who has lived in the Southeastern United States for the majority of his adult life. His experience with disparities in healthcare are derived not from personal experience, but from professional experiences in his roles as an Associate Professor and clinic director at an academic medical center.

## Conflict of Interest Statement

Kiiya Shibata receives a stipend from Vanderbilt University. Dr. Michael de Riesthal, Dr. Ryan Darby, and Jerica Reeder receive a salary from Vanderbilt University Medical Center. Data was derived from the Vanderbilt University Medical Center population.

## IRB Approval

This research was approved by the IRB at Vanderbilt University. Approval number 220484.

## References


Alzheimer's Association. (2018). 2018 Alzheimer's disease facts and figures. *Alzheimer's & Dementia*, *14*(3), 367–429. https://doi.org/10.1016/j.jalz.2018.02.001

Alzheimer's Association. (2019). 2019 Alzheimer's disease facts and figures. *Alzheimer's & Dementia*, *15*(3), 321–387. https://doi.org/10.1016/j.jalz.2019.01.010

Ansaldo, A. I., Marcotte, K., Scherer, L. C., & Raboyeau, G. (2008). Language therapy and bilingual aphasia: Clinical implications of psycholinguistic and neuroimaging research. *Journal of Neurolinguistics*, *21*(6), 539–557. https://doi.org/10.1016/j.jneuroling.2008.02.001

Aranda, M. P., Kremer, I. N., Hinton, L., Zissimopoulos, J., Whitmer, R. A., Hummel, C. H., Trejo, L., & Fabius, C. (2021). Impact of dementia: Health disparities, population trends, care interventions, and economic costs. *Journal of the American Geriatrics Society*, *69*(7), 1774–1783. https://doi.org/10.1111/jgs.17345

Bahia, V. S. (2007). Underdiagnosis of frontotemporal lobar degeneration in Brazil. *Dementia & Neuropsychologia*, *1*(4), 361–365. https://doi.org/10.1590/s1980-57642008dn10400006

Bellinger, J. D., Hassan, R. M., Rivers, P. A., Cheng, Q., Williams, E. M., & Glover, S. H. (2010). Specialty Care Use in US Patients with Chronic Diseases. *International Journal of Environmental Research and Public Health*, *7*(3), 975–990. https://doi.org/10.3390/ijerph7030975

Buchalter, R. B., Gentry, E. G., Willis, M. A., & McGinley, M. P. (2023). Disparities in spatial access to neurological care in Appalachia: a cross-sectional health services analysis. *The Lancet Regional Health. Americas*, *18*, 100415. https://doi.org/10.1016/j.lana.2022.100415

Centers For Disease Control. (n.d.). *Leading causes of death and injury*. CDC Injury Center. https://www.cdc.gov/injury/wisqars/LeadingCauses.html

Cohen, J., Cohen, P., West, S. G., & Aiken, L. S. (2014). Applied Multiple Regression/Correlation Analysis for the Behavioral Sciences. In *Psychology Press eBooks*. https://doi.org/10.4324/9781410606266

Conron, K. J., Luhur, W., & Goldberg, S. K. (2020). *Estimated number of US LGBT adults in Large Metropolitan Statistical Areas (MSA)*. The Williams Institute, UCLA.

Cooper, C., Tandy, A. R., Balamurali, T. B. S., & Livingston, G. (2010). A Systematic Review and Meta-Analysis of Ethnic Differences in Use of Dementia Treatment, care, and Research. *American Journal of Geriatric Psychiatry*, *18*(3), 193–203. https://doi.org/10.1097/jgp.0b013e3181bf9caf

Cousins, E., De Vries, K., & Dening, K. H. (2020). LGBTQ+ people living with dementia: an under-served population. *British Journal of Neuroscience Nursing*. https://doi.org/10.12968/bjnn.2020.16.5.221

Coyle-Gilchrist, I., Dick, K. M., Patterson, K., Rodríquez, P. V., Wehmann, E., Wilcox, A., Lansdall, C. J., Dawson, K., Wiggins, J., Mead, S., Brayne, C., & Rowe, J. B. (2016). Prevalence, characteristics, and survival of frontotemporal lobar degeneration syndromes. *Neurology*, *86*(18), 1736–1743. https://doi.org/10.1212/wnl.0000000000002638

Danciu, I., Cowan, J. D., Basford, M. A., Wang, X., Saip, A., Osgood, S., Shirey-Rice, J. K., Kirby, J., & Harris, P. A. (2014). Secondary use of clinical data: the Vanderbilt approach. *Journal of Biomedical Informatics*. https://doi.org/10.1016/j.jbi.2014.02.003

Di Lorito, C., Bosco, A., Peel, E., Hinchliff, S., Dening, T., Calasanti, T., De Vries, B., Cutler, N., Fredriksen-Goldsen, K. I., & Harwood, R. H. (2021). Are dementia services and support organisations meeting the needs of Lesbian, Gay, Bisexual and Transgender (LGBT) caregivers of LGBT people living with dementia? A scoping review of the literature. *Aging & Mental Health*, *26*(10), 1912–1921. https://doi.org/10.1080/13607863.2021.2008870

Duffy, J. R. (2006). Apraxia of speech in degenerative neurologic disease. *Aphasiology*, *20*(6). https://doi.org/10.1080/02687030600597358

Duran-Aniotz, C., Orellana, P., Leon Rodriguez, T., Henriquez, F., Cabello, V., Aguirre-Pinto, M. F., Escobedo, T., Takada, L. T., Pina-Escudero, S. D., Lopez, O., Yokoyama, J. S., Ibanez, A., Parra, M. A., & Slachevsky, A. (2021). Systematic review: Genetic, neuroimaging, and fluids biomarkers for frontotemporal dementia across latin america countries. *Frontiers in Neurology*, *12*. https://doi.org/10.3389/fneur.2021.663407

Farina, M. P., Hayward, M., Kim, J. H., & Crimmins, E. M. (2020). Racial and Educational Disparities in Dementia and Dementia-Free Life Expectancy. *The Journals of Gerontology: Series B*, *75*(7), e105–e112. https://doi.org/10.1093/geronb/gbz046

Foth, T., & Leibing, A. (2022). Rethinking dementia as a queer way of life and as 'crip possibility': A critique of the concept of person in person-centredness. *Nursing Philosophy*, *23*(1). https://doi.org/10.1111/nup.12373

Franks, P., & Clancy, C. M. (1997). Referrals of adult patients from primary care: demographic disparities and their relationship to HMO insurance. *Journal of Family Practice*, *45*(1), 47–53. https://cdn.mdedge.com/files/s3fs-public/jfp-archived-issues/1997-volume_44-45/JFP_1997-07_v45_i1_referrals-of-adult-patients-from-primary.pdf

Garcia, M. A., Downer, B., Chiu, C., Saenz, J. L., Rote, S., & Wong, R. (2019). Racial/Ethnic and Nativity Differences in Cognitive Life Expectancies Among Older Adults in the United States. *Gerontologist*, *59*(2), 281–289. https://doi.org/10.1093/geront/gnx142

Garcia, O. (2011). From language garden to sustainable languaging: Bilingual education in a global world. *Nabe Perspectives*. https://ofeliagarciadotorg.wordpress.com/wp-content/uploads/2011/02/2012nabenewsletter_34n1_nov2011_dec2011.pdf

Gorno-Tempini, M. L., Hillis, A. E., Weintraub, S., Kertesz, A., Mendez, M., Cappa, S. F., Ogar, J. M., Rohrer, J. D., Black, S., Boeve, B. F., Manes, F., Dronkers, N. F., Vandenberghe, R., Rascovsky, K., Patterson, K., Miller, B. L., Knopman, D. S., Hodges, J. R., Mesulam, M. M., & Grossman, M. (2011). Classification of primary progressive aphasia and its variants. *Neurology*, *76*(11), 1006–1014. https://doi.org/10.1212/wnl.0b013e31821103e6

Guimarães, H. C., Vale, T. C., Pimentel, V., De Sá, N. C., Beato, R. G., & Caramelli, P. (2013). Analysis of a case series of behavioral variant frontotemporal dementia: Emphasis on diagnostic delay. *Dementia & Neuropsychologia*, *7*(1), 55–59. https://doi.org/10.1590/s1980-57642013dn70100009

Guo, Y. G., Li, Q., Yang, X., Jaffee, M. S., Wu, Y., Wang, F., & Bian, J. (2022). Prevalence of Alzheimer's and Related Dementia Diseases and Risk Factors Among Transgender Adults, Florida, 2012–2020. *American Journal of Public Health*, *112*(5), 754–757. https://doi.org/10.2105/ajph.2022.306720

Harper, P. M. (2019). How healthcare professionals can support older LGBTQ+ people living with dementia. *Nursing Older People*, *31*(5), 16–21. https://doi.org/10.7748/nop.2019.e1115

Hayes-Larson, E., Mobley, T. M., Mungas, D. M., Seamans, M. J., Glymour, M. M., Gilsanz, P., DeCarli, C., Whitmer, R. A., & Mayeda, E. R. (2022). Accounting for lack of representation in dementia research: Generalizing KHANDLE study findings on the prevalence of cognitive impairment to the California older population. *Alzheimers & Dementia*, *18*(11), 2209–2217. https://doi.org/10.1002/alz.12522

Hogan, D. B., Jette, N., Fiest, K. M., Roberts, J. I., Pearson, D., Smith, E. E., Roach, P., Kirk, A. G., Pringsheim, T., & Maxwell, C. J. (2016). The Prevalence and Incidence of Frontotemporal Dementia: a Systematic Review. *Canadian Journal of Neurological Sciences*, *43*(S1), S96–S109. https://doi.org/10.1017/cjn.2016.25

*Human right campaign tennessee scorecard*. (n.d.). Human Rights Campaign. https://www.hrc.org/resources/state-scorecards/tennessee-3

*Interactive: The Unequal States of America*. (n.d.). Economic Policy Institute. https://www.epi.org/multimedia/unequal-states-of-america/

Jutkowitz, E., Kane, R. L., Gaugler, J. E., MacLehose, R. F., Dowd, B. E., & Kuntz, K. M. (2017). Societal and Family Lifetime Cost of Dementia: Implications for Policy. *Journal of the American Geriatrics Society*, *65*(10), 2169–2175. https://doi.org/10.1111/jgs.15043

Kawakatsu, S., Kobayashi, R., Hayashi, H., Morioka, D., Utsunomiya, A., Kabasawa, T., Ohe, R., & Otani, K. (2021). Clinicopathological heterogeneity of Alzheimer's disease with pure Alzheimer's disease pathology: Cases associated with dementia with Lewy bodies, very early-onset dementia, and primary progressive aphasia. *Neuropathology*, *41*(6), 427–449. https://doi.org/10.1111/neup.12765

Kelley, A. S., McGarry, K., Gorges, R. J., & Skinner, J. (2015). The Burden of Health Care Costs for Patients With Dementia in the Last 5 Years of Life. *Annals of Internal Medicine*, *163*(10), 729–736. https://doi.org/10.7326/m15-0381

Kendi, I. X. (2019). *How to be an antiracist*. One World.

Kim, E. J. K., Moon, S. Y. M., Kim, H. J. K., Jung, N.-Y., Lee, S. M. L., & Kim, Y. E. K. (2022). Semantic variant primary progressive aphasia with a pathogenic variant p.Asp40Gly in the ANXA11 gene. *European Journal of Neurology*, *29*(10), 3124–3126. https://doi.org/10.1111/ene.15455

King, A. J. (2021). Queer futures? Forget it! Dementia, queer theory and the limits of normativity. *Journal of Aging Studies*, *63*, 100993. https://doi.org/10.1016/j.jaging.2021.100993

Knopman, D. S., & Roberts, R. O. (2011). Estimating the number of persons with frontotemporal lobar degeneration in the US population. *Journal of Molecular Neuroscience*, *45*(3), 330–335. https://doi.org/10.1007/s12031-011-9538-y

Lopez, M., Krogstad, J., & Passel, J. (2023, September 8). *Who is hispanic?* Pew Research Center. https://www.pewresearch.org/short-reads/2023/09/05/who-is-hispanic/#:~:text=In%201976%2C%20the%20U.S.%20Congress,of%20Spanish%20origin%20or%20descent.%E2%80%9D

Majounie, E., Renton, A. E., Mok, K. Y., Dopper, E. G., Waite, A. J., Rollinson, S., Chiò, A., Restagno, G., Nicolaou, N., Simón-Sánchez, J., Van Swieten, J. C., Abramzon, Y., Johnson, J. O., Sendtner, M., Pamphlett, R., Orrell, R. W., Mead, S., Sidle, K., Houlden, H., & Traynor, B. J. (2012). Frequency of the C9orf72 hexanucleotide repeat expansion in patients with amyotrophic lateral sclerosis and frontotemporal dementia: a cross-sectional study. *Lancet Neurology*, *11*(4), 323–330. https://doi.org/10.1016/s1474-4422(12)70043-1

Malcolm, T., Lerman, A., Korytkowska, M., Vonk, J. M. J. V., & Obler, L. K. O. (2019). Primary progressive aphasias in bilinguals and multilinguals. In J. W. S. Schwieter & M. Paradis (Eds.), *The Handbook of the Neuroscience of Multilingualism*. https://doi.org/10.1002/9781119387725.ch28

Matthews, K. A., Xu, W., Gaglioti, A. H., Holt, J. B., Croft, J. B., Mack, D., & McGuire, L. C. (2018). Racial and ethnic estimates of Alzheimer's disease and related dementias in the United States (2015–2060) in adults aged ≥65 years. *Alzheimers & Dementia*, *15*(1), 17–24. https://doi.org/10.1016/j.jalz.2018.06.3063

Mayeda, E. R. (2019). Invited Commentary: Examining Sex/Gender Differences in Risk of Alzheimer Disease and Related Dementias—Challenges and Future Directions. *American Journal of Epidemiology*, *188*(7), 1224–1227. https://doi.org/10.1093/aje/kwz047

Mayeda, E. R., Glymour, M. M., Quesenberry, C. P., & Whitmer, R. A. (2016). Inequalities in dementia incidence between six racial and ethnic groups over 14 years. *Alzheimers & Dementia*, *12*(3), 216–224. https://doi.org/10.1016/j.jalz.2015.12.007

Mehta, K. M., & Yeo, G. (2016). Systematic review of dementia prevalence and incidence in United States race/ethnic populations. *Alzheimers & Dementia*, *13*(1), 72–83. https://doi.org/10.1016/j.jalz.2016.06.2360

Mendez, M. F., Chavez, D., & Akhlaghipour, G. (2020). Bilingualism delays expression of Alzheimer's clinical syndrome. *Dementia and Geriatric Cognitive Disorders*, *48*(5–6). https://doi.org/10.1159/000505872

Mesulam, M. M. (2001). Primary progressive aphasia. *Annals of Neurology*, *49*(4), 425–432. https://doi.org/10.1002/ana.91

Mesulam, M. M., Coventry, C., Bigio, E. H., Geula, C., Thompson, C. K., Bonakdarpour, B., Gefen, T., Rogalski, E., & Weintraub, S. (2021). *Nosology of Primary Progressive Aphasia and the Neuropathology of Language* (pp. 33–49). Springer International Publishing eBooks. https://doi.org/10.1007/978-3-030-51140-1_3

Miner, A. (2022). Brain Barriers: Elucidating Complex Language-Based Barriers to Neurological Care in Limited-English Proficiency, Refugee, and Immigrant Populations (P17-13.003). *Neurology*, *98*(18_supplement). https://doi.org/10.1212/wnl.98.18_supplement.1445

Moon, H., Badana, A. N., Hwang, S., Sears, J., & Haley, W. E. (2019). Dementia Prevalence in Older Adults: Variation by Race/Ethnicity and Immigrant Status. *American Journal of Geriatric Psychiatry*, *27*(3), 241–250. https://doi.org/10.1016/j.jagp.2018.11.003

Ng, T. P., Niti, M., Chiam, P., & Kua, E. H. (2007). Ethnic and Educational Differences in Cognitive Test Performance on Mini-Mental State Examination in Asians. *American Journal of Geriatric Psychiatry*, *15*(2), 130–139. https://doi.org/10.1097/01.jgp.0000235710.17450.9a

Onyike, C. U., Shinagawa, S., & Ellajosyula, R. (2020). *Frontotemporal Dementia: A Cross-Cultural Perspective* (pp. 141–150). Springer International Publishing eBooks. https://doi.org/10.1007/978-3-030-51140-1_10

Pan, S., Stutzbach, J., Reichwein, S., Lee, B. R., & Dahodwala, N. (2014). Knowledge and Attitudes About Parkinson's Disease Among a Diverse Group of Older Adults. *Journal of Cross-Cultural Gerontology*, *29*(3), 339–352. https://doi.org/10.1007/s10823-014-9233-x

Petersen, J. D., Wehberg, S., Packness, A., Svensson, N. H., Hyldig, N., Raunsgaard, S., Andersen, M. K. K., Ryg, J., Mercer, S. W., Søndergaard, J., & Waldorff, F. B. (2021). Association of Socioeconomic Status With Dementia Diagnosis Among Older Adults in Denmark. *JAMA Network Open*, *4*(5), e2110432. https://doi.org/10.1001/jamanetworkopen.2021.10432

Pijnenburg, Y. A., Gillissen, F., Jonker, C., & Scheltens, P. (2004). Initial complaints in frontotemporal lobar degeneration. *Dementia and Geriatric Cognitive Disorders*, *17*(4), 302–306. https://doi.org/10.1159/000077159

Qian, W., Schweizer, T. A., & Fischer, C. E. (2014). Impact of socioeconomic status on initial clinical presentation to a memory disorders clinic. *International Psychogeriatrics*, *26*(4), 597–603. https://doi.org/10.1017/s1041610213002299

R Core Team. (2021). *R: A language and environment for statistical computing (2023.06.1+524)* [Software]. R Foundation for Statistical Computing. https://www.R-project.org/

Roden, D., Pulley, J., Basford, M., Bernard, G., Clayton, E. W., Balser, J., & Masys. (2008). Development of a Large-Scale De-Identified DNA Biobank to enable personalized Medicine. *Clinical Pharmacology & Therapeutics*, *84*(3), 362–369. https://doi.org/10.1038/clpt.2008.89

Saadi, A., Himmelstein, D. U., Woolhandler, S., & Mejia, N. I. (2017). Racial disparities in neurologic health care access and utilization in the United States. *Neurology*, *88*(24), 2268–2275. https://doi.org/10.1212/wnl.0000000000004025

Sahadevan, S., Saw, S. M., Gao, W., Tan, L. C., Chin, J. J., Hong, C. Y., & Venketasubramanian, N. (2008). Ethnic Differences in Singapore's Dementia Prevalence: The Stroke, Parkinson's Disease, Epilepsy, and Dementia in Singapore study. *Journal of the American Geriatrics Society*, *56*(11), 2061–2068. https://doi.org/10.1111/j.1532-5415.2008.01992.x

Schulz, R., & Beach, S. R. (1999). Caregiving as a Risk Factor for Mortality. *JAMA*, *282*(23), 2215. https://doi.org/10.1001/jama.282.23.2215

Sentell, T., Valcour, N., Ahn, H. S., Miyamura, J., Nakamoto, B. K., Chow, D. C., Masaki, K., Seto, T. B., Chen, J. J., & Shikuma, C. M. (2015). High Rates of Native Hawaiian and Older Japanese Adults Hospitalized with Dementia in Hawai'i. *Journal of the American Geriatrics Society*, *63*(1), 158–164. https://doi.org/10.1111/jgs.13182

Shah, B. R., Khan, N. A., O'Donnell, M., & Kapral, M. K. (2015). Impact of Language Barriers on Stroke Care and Outcomes. *Stroke*, *46*(3), 813–818. https://doi.org/10.1161/strokeaha.114.007929

Shamsi, H. A., Almutairi, A. G., Mashrafi, S. S. A., & Kalbani, T. S. A. (2020). Implications of Language Barriers for Healthcare: A Systematic Review. *Oman Medical Journal*, *35*(2), e122. https://doi.org/10.5001/omj.2020.40

The Association for Frontotemporal Lobar Degeneration. (2022). *Research and Medical Centers | AFTD*. AFTD. https://www.theaftd.org/what-is-ftd/research-and-medical-centers/

Umemoto, K. (1989). "On Strike!" San Francisco State College Strike, 1968–69: The Role of Asian American Students. *Amerasia Journal*, *15*(1), 3–41. https://doi.org/10.17953/amer.15.1.7213030j5644rx25

US Census Bureau. (n.d.). *U.S. Census Bureau QuickFacts: Tennessee; United States*. Census Bureau QuickFacts. https://www.census.gov/quickfacts/fact/table/TN,US/PST045222

US Census Bureau. (2022a, June 9). *Income and Poverty in the United States: 2020*. Census.gov. https://www.census.gov/library/publications/2021/demo/p60-273.html

US Census Bureau. (2022b, December 13). *American community survey 2021*. Census.gov. https://data.census.gov/table/ACSDP5Y2021.DP02

Vanderbilt University. (2023, September 12). *Expanded data collection provides a more complete patient story and helps Vanderbilt Health advance health equity*. Vanderbilt University. https://news.vumc.org/2023/09/12/expanded-data-collection-provides-a-more-complete-patient-story-and-helps-vanderbilt-health-advance-health-equity/

VUMC Office of Government and Community Affairs. (n.d.). *County by county impact*. VUMC.org. https://www.vumc.org/ogca/county-county-impact

Warren, L. E., Shi, Q., Young, K., Borenstein, A. R., & Martiniuk, A. (2015). Prevalence and incidence of dementia among indigenous populations: a systematic review. *International Psychogeriatrics*, *27*(12), 1959–1970. https://doi.org/10.1017/s1041610215000861

Xu, J. X., Zia, Y. X., Meng, M. M., Liu, F. L., Che, P. C., Zhang, Y. Z., Wang, Y. W., Cai, L. C., Qin, W. Q., & Zhang, N. Z. (2023). Clinical features and biomarkers of semantic variant primary progressive aphasia with MAPT mutation. *Alzheimer's Research & Therapy*, *15*(1), 1–14. https://doi.org/10.1186/s13195-023-01176-y

Zinn, H. (1980). *A people's history of the United States*. HarperCollins Publishers.