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A Systematic Review on the Evidence of Misdiagnosis in Dementia and Its Impact on Accessing Dementia Care

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ABSTRACT

Background: Whilst there is a drive to increase diagnosis rates in dementia, there is a lack of attention on getting a correct and timely subtype diagnosis. For people with a rarer subtype of dementia, getting the correct diagnosis, and subsequent care, might be more difficult than for people aged 65+ presenting with the more common symptoms of Alzheimer's disease dementia. Thus, the aim of this mixed-method systematic review was to synthesise the evidence base on misdiagnosis of dementia.

Methods: Misdiagnosis in dementia was defined as either receiving an initial incorrect dementia subtype diagnosis or receiving an incorrect non-dementia diagnosis. Post-mortem assessments of subtype diagnosis were excluded. Nine databases were searched in June 2023, with screening of titles and abstracts and consequent full texts completed independently by two researchers. Findings were synthesised using narrative synthesis.

Results: Twenty studies were included. Studies were categorised into four themes: (i) Factors associated with delayed diagnosis or misdiagnosis; (ii) Difficulties related to the diagnostic process; (iii) Economic consequences of misdiagnosis; and (iv) Experiences of delayed diagnosis or help-seeking. People with Lewy Body dementia or behavioural variant fronto-temporal dementia were found to experience longer diagnosis times and often incorrect initial diagnoses. Whilst evidence is limited regarding the economic impacts, evidence from the US points towards increased economic costs of misdiagnosis.

Conclusions: There is an urgent need to investigate the rates and emotional and economic impacts of misdiagnosis on people with dementia, their carers, and the health and social care system. Advancing the evidence base is crucial to reduce misdiagnosis and inform clinical practice.

Summary

- Evidence focussing on all-cause dementia and specific subtypes of dementia indicates increased misdiagnosis and delayed diagnosis rates for people with rarer subtypes of dementia.
- There is no evidence to date on cross-country comparisons of misdiagnosis rates for different dementia subtypes.
- Limited evidence exists on the economic impacts of misdiagnosis, focussing on the healthcare system only and not on the individual.

1 | Introduction

Globally, over 55 million people are living with dementia [1]. According to National Institute for Excellence (NICE) guidance [2], 40% of people with dementia (1 million overall) in the UK have a rarer subtype of dementia that is not associated with Alzheimer's disease. These include Lewy Body, vascular, Parkinson's disease, behavioural-variant fronto-temporal (FTD), and semantic dementia, as well as progressive non-fluent aphasia (PNFA). People can also experience a mixed dementia, which can be a mix of any different subtypes.

Each subtype is characterised by different symptom profiles. Lewy Body and Parkinson's disease dementia for example present with motor difficulties, and the former can also include more hallucinations [3]. Behavioural-variant FTD is characterised by more pronounced behavioural difficulties, such as agitation, apathy, and sleep difficulties, whereas language-led dementias such as semantic dementia and PNFA present with language difficulties as the leading symptom [4, 5]. These symptom profiles are associated with different areas of brain atrophy [6], and can results in different difficulties in initiating and performing everyday activities such as managing finances and medication, preparing a hot meal, and dressing [7].

Living with a rare dementia is often linked to increased barriers in accessing social care and support services [8, 9]. People living with Lewy Body dementia and their unpaid carers often do not receive sufficient information and support following diagnosis [8]. However, receiving the correct diagnosis in the first place can be difficult for people with rarer dementias. Evidence suggests that people can wait several years to receive a correct subtype diagnosis, with some not receiving an accurate diagnosis until post-mortem meaning that many people living with dementia may live their entire dementia journey with an incorrect diagnosis (i.e., [10–13]). This can be caused by a variety of factors, including reduced health care professional knowledge of dementia subtypes and age (young-onset dementia), leading to some people with dementia facing the dementia journey without certainty and adequate support. Yet, to date there has been no systematic synthesis of the evidence of misdiagnosis of dementia subtypes and the impact on accessing support.

Therefore, the aim of this mixed-method systematic review was to explore the evidence on misdiagnosis of dementia and its potential impact on people with dementia and their unpaid carers. For the purposes of this review a misdiagnosis was defined as either receiving an initial incorrect dementia subtype diagnosis, or receiving an incorrect non-dementia diagnosis. Improving the accuracy of a dementia diagnosis is vital to ensuring that people with dementia and their families receive the right pharmacological and/or non-pharmacological support and information to prepare for the dementia journey.

2 | Methods

This systematic review has been registered on PROSPERO before formal searches were conducted [Ref: CRD42023426874].

2.1 | Population

This systematic review focussed on people living with dementia with a diagnosis, and on unpaid carers of people with a diagnosis of dementia. No restrictions were placed on living location, age, or subtype diagnosis. For the purposes of this review unpaid carers had to be aged 18+.

2.2 | Inclusion/Exclusion Criteria

Only research studies were included which provided new evidence. No reviews, letters, editorials, commentaries, or conference abstracts were included. Studies published from 2010 onwards in English, German, Italian, and Spanish were included. 2010 was chosen as a cut-off date for earliest publications because of introductions and implementations of National Dementia Strategies across different countries and foci on targeted diagnosis rates of dementia. Given the lack of evidence synthesis in the field, this review included quantitative, qualitative, and mixed-methods studies to generate a clear overview and understanding of the state of research on misdiagnosis in dementia.

2.3 | Search Strategy

A search strategy was developed by the research team, which included search terms to capture the following dimensions: dementia and its subtypes; misdiagnosis; and inequality in or barriers to diagnosis.

The search was developed and piloted using the Ovid search engine. After the research team agreed on the search terms to be used, one reviewer with experience in designing and conducting systematic reviews (WSR) performed the final search in June 2023. Through Ovid, the search was simultaneously run in the following electronic databases: Journals@Ovid full text; Your Journals@Ovid; APA PsycArticles Full Text; Embase; Global Health; HMIC Health Management Information Consortium; Ovid MEDLINE; and APA Psycinfo. On the same day, the search was replicated at Web of Science, to include the Web of Science Core Collection database. A complete list of search terms and the search strategy developed at Ovid are provided in Appendix 1.

2.4 | Study Selection

Two researchers assessed title and abstracts of retrieved records against inclusion criteria and excluded articles that failed to meet inclusion criteria in Stage 1. The task was shared among six researchers, ensuring that each title was assessed by two reviewers (C.T., K.H., C.G., W.S.R., B.H., J.W., A.C., A.V., I.C.). The selected full text records were read in Stage 2 by two researchers again, and articles that met the inclusion criteria were included in the review. The task was split among the research team again. Any discrepancies at stage 1 or 2 were resolved in discussion with the wider research team.

2.5 | Data Extraction and Synthesis

Data from the included studies were extracted by two research team members (J.W., W.S.R.) into an Excel sheet, by extracting key study characteristics and results. Considering the heterogeneity of studies, the extraction for was piloted by extracting data from one qualitative and one quantitative study and revised by the extractors iteratively.

Data were narratively synthesised by four research team members (W.S.R., C.T., C.G., J.W.). This process involves extracting the specific findings when a meta-analysis may not be feasible due to wide-ranging differences in methodological procedures or outcome variables across included studies [14]. There was no minimum number of studies to be included. The collation of the evidence involved narratively synthesising the data extracted, and clustering the included studies together based on their focus of misdiagnosis, as defined by the subheadings included in the results section. Both quantitative and qualitative findings were summarised and synthesised.

2.6 | Assessment of Study Bias

We assessed the quality of each included paper, using the CASP for qualitative studies (2018) and the Risk of Bias in Non-randomized Studies—of Exposure (ROBINS-E) [15] for quantitative studies. For the ROBINS-E tool, total scores can range from 0 to 6 with higher scores meaning higher probability of risk of bias. Quality assessments were performed by two researchers independently. Any discrepancies between ratings were discussed jointly. If unclear after discussion, a third researcher was consulted. Quality ratings did not influence study selection, but will be used in guiding the discussion of findings and drawing conclusions.

3 | Results

3.1 | Overview of Included Studies

Figure 1 shows the PRISMA flowchart of included studies. Table 1 shows the descriptive characteristics of the 20 research studies which met the inclusion criteria for this systematic review. The majority ($n = 16$) were quantitative (9 cross-sectional studies, 4 cohort studies, 2 case-control studies and 1 case series) and five were qualitative. There was a total of 12 countries in which these studies were conducted, with some being conducted in more than one country. Nearly all the studies were conducted with populations in North America (10 in the USA and two in Canada) and Europe (five in the Netherlands, one in England, one each in Germany, Slovenia, Spain, Sweden and the UK). One study was conducted respectively in Australia, Brazil and Japan. All included studies were published in English.

Several of the studies focussed on populations with a diagnosis of any dementia ($n = 8$). However, several focussed on populations diagnosed with specific dementia subtypes; frontotemporal dementia (FTD, $n = 1$), Alzheimer's Disease ($n = 1$), or specified early-/young-onset dementia ($n = 1$) and Lewy Body Dementia ($n = 3$).

Studies were categorised into (i) Factors associated with delayed diagnosis or misdiagnosis, including inadequate assessments ($n = 10$); (ii) Difficulties related to the diagnostic process ($n = 6$); (iii) Economic consequences of misdiagnosis ($n = 2$); and (iv) Experiences of obtaining a dementia diagnosis ($n = 4$).

3.2 | Factors Associated With Delayed Diagnosis and Misdiagnosis

Studies employed different designs to assess potential predictors of delayed diagnosis and misdiagnosis of dementia, including case series [16], cross-sectional [17–21] case-control [12, 22], and cohort studies [10, 13, 23].

Among demographic characteristics, ethnicity was the factor most commonly found to be associated with delayed diagnosis and misdiagnosis. Lin et at. (2021) found that among adults with cognitive functional decline consistent with dementia in the US, a higher proportion of non-Hispanic Blacks and Hispanics had a missed or delayed clinical diagnosis of dementia, as compared to non-Hispanic Whites. They also noted that, by the time of receiving a dementia diagnosis, Hispanic and Black people with dementia had poorer cognitive function and more functional limitations than those from White ethnic backgrounds.

Age was another potential predictor reported in some studies, but with conflicting results. Whilst [18] found that, in the US, increased patient age was associated with increased odds of having a delayed dementia diagnosis (over 1 year after symptoms onset), another study in the US [13] reported that increased aged was associated with increased likelihood of false positive dementia diagnosis in Medicare registers.

Studies also assessed clinical characteristics associated with delayed diagnosis and misdiagnosis of dementia. Worse cognitive status was associated with a false positive diagnosis of dementia in the US's Medicare registers [13]. Conversely, another US study [21] found that patients with better overall cognition and milder disease severity were more likely to be diagnosed by non-specialist physicians as false positive cases of behavioural variant frontotemporal dementia.

Three studies found that other psychiatric conditions, particularly depression and mood disorders, seemed to interfere with

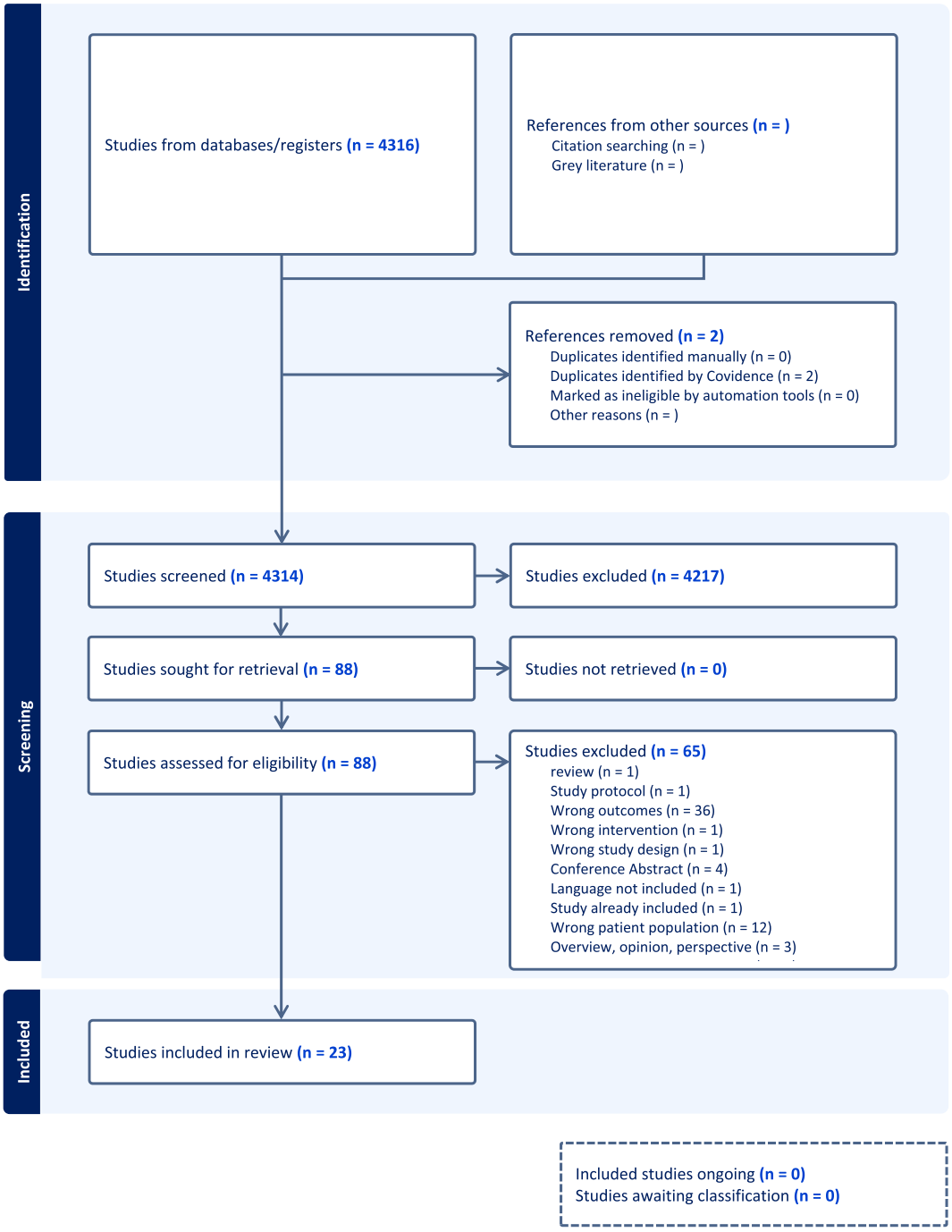

FIGURE 1 | PRISMA flowchart.

dementia diagnosis. Whilst [18] found that mood symptoms were associated with increased likelihood of initially receiving an incorrect diagnosis among patients with frontotemporal dementia, Shinagawa et al. [21] found that patients with a false positive diagnosis of behavioural variant frontotemporal dementia were more likely to be depressed. In the study conducted by Draper et al. [19], depression was found to be associated with increased time to dementia diagnosis from symptom onset.

Based on the assumption that cognitive screening is an important step towards timely diagnosis of dementia, one study assessed the proportion of older people in a Japanese city undergoing dementia screening [17]. Twelve percent of participants reported they had previously been screened for dementia in local health services. Among participants who screened positive for possible dementia in the survey, 28.7% reported undergoing a previous dementia screening. Another study, carried out in the US [23], investigated whether patients with a diagnosis of dementia recorded in their electronic health records had also a cognitive measure previously documented. The study found that, among patients with a diagnosis of dementia of any type, 11% had cognitive measures documented in their records in the previous 5 years; among patients with a diagnosis of Alzheimer's disease, 23.6% had cognitive measures documented in their records.

TABLE 1 | Basic characteristics of included studies.

Author	Year	Country	Study type	Study design	Dementia subtypes	Total sample	Study aim/objective	Main outcome(s)/themes	Factors of inequalities
Aihara	2020	Japan	Quantitative	Cross-sectional	Not reported	276	Identify factors that promote public intention to undergo dementia screening	Willingness to undergo dementia screening	Age; Gender; Education; Living status; comorbidities; health status
Barker	2023	USA; Canada; UK	Quantitative	Cross-sectional	Frontotemporal dementia (FTD): Self-reported diagnosis of bvFTD or Pick's Disease, diagnosed PPA, diagnosed PSP/CBS, FTD with ALS, and care partners within each subgroup	655	Chronicling the diagnostic journey, symptoms, and the impact of FTD on distress, quality of life, and independence, in the mild to moderate stages of FTD	Prior "different diagnosis"; number of doctors consulted before FTD diagnosis	FTD subtypes
Beber	2013	Brazil	Quantitative	Case-control	FTD; Alzheimer's Disease	29	Analyse variables associated with misdiagnosis in a group of patients with FTD, Alzheimer's disease (AD), and without neurodegenerative disorders (WND).	Diagnosis prior to evaluation at specialised clinic	Age; Gender; Dementia subtype
Besser	2020	USA	Quantitative	Cross-sectional	Frontotemporal dementia (FTD)	698	Describe the experience of obtaining a diagnosis of frontotemporal degeneration (FTD) for patients and caregivers	Time to diagnosis after symptom onset; initial non-FTD diagnosis	Patient age; caregiver age; patient gender; caregiver gender; patient education level; caregiver education level; relationship of caregiver; caregiver urban/rural residence; caregiver residential status

(Continues)

TABLE 1 | (Continued)

Author	Year	Country	Study type	Study design	Dementia subtypes	Total sample	Study aim/objective	Main outcome(s)/themes	Factors of inequalities
Drabo	2019	USA	Quantitative	Cohort	Multiple subtypes	226,604	Quantify types of physicians diagnosing dementia, subtype diagnoses initially provided and change in diagnoses over time. Access to dementia specialty care, and demographic variation	Incidence of dementia; types of diagnosing professionals; use of specialized dementia care	Healthcare visits; ethnicity; Gender; Dementia subtype
Draper	2016	Australia	Quantitative	Cross-sectional	Young-onset: Alzheimer's Disease, FTD, vascular, PSP, Huntington's Disease, mixed Alzheimer's/ Vascular, Dementia with Lewy bodies, alcohol-related, mixed Alzheimer/Alcohol-related, unknown	88	Identify factors determining time to diagnosis for young-onset dementia (YOD)	Time from symptom onset to first consultation; time from first consultation to diagnosis; time from symptom onset to family awareness of dementia diagnosis	Age; Gender; Education
Hoppe	2019	Netherlands	Qualitative	SSI	Early-onset Dementia	46	Provide better understanding of uncertainties experienced in accessing diagnosis in early-onset dementia	Accepting and maintaining uncertainty; finding explanations; taking action	N/A
Hunter	2015	USA	Quantitative	Cross-sectional	Alzheimer's Disease, vascular dementia, Parkinson's disease	19,543	Assess potentially avoidable medical service utilization and the resulting economic benefits of timely rule-out of AD among US medicare beneficiaries eventually diagnosed with VD or PD	Medical resource use attributable to misdiagnosis of AD	Age; Gender

(Continues)

TABLE 1 | (Continued)

Author	Year	Country	Study type	Study design	Dementia subtypes	Total sample	Study aim/objective	Main outcome(s)/themes	Factors of inequalities
Lin	2021	USA	Quantitative	Cohort	All dementia	3966	Examine racial and ethnic disparities in the timeliness of receiving a clinical diagnosis of dementia	Missed or delayed diagnosis	Ethnicity; age; Gender; ADL & IADL limitations; comorbidities; residence; education; dementia severity; insurance type
Maserejian	2020	USA	Quantitative	Cohort	All dementia; Alzheimer's disease	141,328	Describe the frequency and factors associated with documented cognitive measures among patients prior to the diagnosis of AD or dementia in a large U.S. EHR database	Proportion of patients with prior [to diagnosis] cognitive measure	Age; Gender; ethnicity; household income; medical insurance type; inpatient admissions' encounters with different healthcare types; medication prescriptions
Mendez	2013	USA	Quantitative	Cross-sectional	bvFTD	95	Characterise presenting symptoms of patients clinically diagnosed with behavioural variant frontotemporal dementia and who had different autopsy neuropathologic findings	Cognition, behaviour and motor symptoms	Dementia subgroups?
Novek	2021	Canada	Qualitative	SSI	Young-onset Dementia	6	Critically examine the process of accessing and delivering a diagnosis of young onset dementia from the perspectives of people living with young onset dementia, family members, and providers	Identification; navigation [through services]; services' permeability; appearances and adjudications	N/A
Sannemann	2011	Germany, Spain, Netherlands, Sweden, Slovenia	Quantitative	Cross-sectional	Alzheimer's Disease	343	Provide an up-to-date overview on GPs' opinions on early and pre-dementia diagnosis for AD and gain insight into diagnostic processes	General practitioners' attitudes towards dementia diagnosis	Country

(Continues)

TABLE 1 | (Continued)

Author	Year	Country	Study type	Study design	Dementia subtypes	Total sample	Study aim/objective	Main outcome(s)/themes	Factors of inequalities
Schrauf	2011	USA	Quantitative	Cross-sectional	All dementia	42	Characterise the duration and direction of long versus short pathways to diagnosis among African American and Hispanics	Median times-to-diagnosis	Age; Gender; ethnicity
Shinagawa	2016	USA	Quantitative	Cross-sectional	bvFTD	4497	Examine the accuracy of community clinicians' diagnoses of bvFTD and to identify patient characteristics leading to misdiagnosis	False-positive diagnosis	Age; Gender; initial diagnosis
Surendranathan	2020	UK	Quantitative	Case-control	Lewy Bodies; non-Lewy Bodies	219	Investigate clinical diagnostic pathways of patients with Lewy body dementia to assess if difficulties in diagnosis may be contributing to these differences	Time to diagnosis; number of prior [wrong] diagnoses; number of clinical assessments/ appointments before final diagnosis	Type of dementia (DLB vs. non-DLB)
Sutcliffe	2015	England	Qualitative	Focus groups	All Dementia	27	Present views of people with dementia and carers on: positive and negative experiences of dementia care; access to information and its communication; suggestions to improve dementia care	Experiences [about diagnosis and support]; care and service receipt; information and communication; suggested improvement to care	N/A
Van Vliet	2011	Netherlands	Qualitative	SSI	Young-onset Dementia	92	Investigate the barriers to diagnosis and to develop a typology of the diagnosis pathway for early-onset dementia caregivers	Behavioural changes; impact [of symptoms] on family life; misattribution of symptoms; help-seeking; hiding problems and denial; non-responsiveness, misdiagnosis and inadequate help	N/A

(Continues)

TABLE 1 | (Continued)

Author	Year	Country	Study type	Study design	Dementia subtypes	Total sample	Study aim/objective	Main outcome(s)/themes	Factors of inequalities
Vergouw	2020	Netherlands	Quantitative	Case study	Dementia with Lewy Bodies mimics	22	Present clinicopathologic series of false-positive Lewy Body Dementia cases	Clinical characteristics	N/A
Zhu	2019	USA	Quantitative	Cohort	All dementia	495	Examine how misidentification of dementia affects estimation of medicare costs in a largely minority cohort of participants for whom accurate in-person diagnoses are available	Misclassification of dementia (false negative and false positive)	Age; Gender; ethnicity; marital status; Education; months of medical insurance; comorbidities

3.3 | Difficulties Related to the Diagnostic Process

Six studies sought to evaluate differential experiences of the diagnostic pathway in dementia. Studies examined factors associated with variation in timely diagnosis [18], impact of behavioural symptoms [12], or clinicians' knowledge [24, 25] on likelihood of correct diagnosis, and variation in initial diagnosis [26, 27]. Several studies focussed on people living with a form of frontotemporal dementia (FTD) [18]. noted that increasing age, having a spousal caregiver, rural residence, and presence of specific symptoms were associated with increased likelihood of receiving a correct FTD diagnosis within a year of initial symptoms. Similarly, Barker et al. [26] highlighted that among people to receive an eventual FTD diagnosis, half (50%) previously received a different diagnosis, and almost half (49%) had seen at least three doctors before receiving their FTD diagnosis [22]. report similar findings from Brazil, with 90% of people with FTD and 80% with ADD in their study experiencing a misdiagnosis before their final subtype diagnosis.

Two studies investigated the involvement of healthcare practitioners in diagnosis. GPs' views of diagnosis and treatment of Alzheimer's Disease were explored across five European countries [25]. GPs who valued early diagnoses were more likely to see the benefits of pharmacological treatments, but fewer GPs who valued early diagnosis would make changes to their initial diagnosis. Drabo et al. [24] screened electronic health records (EHR) to identify whether people from different socio-demographic groups with dementia come into contact with different groups of medical doctors. Also using EHR, Surendranathan et al. [12] compared healthcare use between those diagnosed with Lewy Body Dementia (LBD), Parkinson's disease dementia (PDD), and other forms of dementia in England. In the 1.4 years prior to a PDD diagnosis, patients were recorded as demonstrating difficulties in activities of daily living due to cognitive impairment (46%), and broad cognitive impairment across multiple domains (57%), with some already receiving pharmacological interventions (39%). The study also found that people who went on to receive a final diagnosis of LBD took significantly longer to receive their diagnosis than those who received a non-LBD diagnosis. Those who eventually received a LBD diagnosis also had more imaging tests, clinical assessments and other dementia diagnoses before their LBD diagnosis.

3.4 | Economic Consequences of Misdiagnosis

Two quantitative studies explored the economic impacts of a misdiagnosis. Hunter et al. [28] compared people previously misdiagnosed with ADD and subsequently correctly diagnosed with vascular Dementia (vD) or Parkinson's disease (PD) to people correctly diagnosed with vD/PD from the beginning. People with a formerly incorrect diagnosis of ADD incurred significantly higher costs every year until their correct diagnosis. Specifically, their annual Medicare costs averaged between $9500-$14,000 US dollars, with medical costs only aligning with those of correctly diagnosed people from the beginning after they had received their accurate diagnosis. The

additional costs comprised of extra inpatient days, emergency room visits, outpatient visits, skilled nursing facility visits, home health care, and equipment. Also utilising data from the US Medicare system, Zhu et al. [13] estimated the effects of dementia misdiagnosis on Medicare expenditures. Only half of the diagnosed cases of dementia were correctly identified by Medicare claims, whilst those with a Medicare identified dementia were costing on average $3487 per person more per year than those with a clinical diagnosis of dementia. Whilst evidence is limited in number and country and thus healthcare system, false identification of dementia can lead to increased economic costs.

3.5 | Experiences of Obtaining a Dementia Diagnosis

Four qualitative studies reported on lived experiences of receiving incorrect diagnoses before being diagnosed with dementia. Three of these studies focussed on young-onset dementia (YOD), collectively showing a failure among healthcare professionals to recognise early signs and symptoms [29–31]. Participants across these studies reported that early symptoms of dementia were frequently not recognised or misattributed to other factors such as burnout, menopause, or alcohol consumption. Healthcare providers often dismissed dementia as unlikely due to the patient's relatively young age (e.g., [29]). However, service providers in Canada also acknowledged the challenges associated with atypical presentations of dementia and the limitations of diagnostic tests [30].

Across these four studies, there was a recurring tendency for healthcare providers to attribute initial symptoms of dementia to mental health difficulties such as depression [32, 33]. For example, in the Netherlands [31], found 34% of carers reported their family members initially received an erroneous diagnosis, while verification of medical records showed 45% received a different diagnosis before being diagnosed with dementia, with mental health diagnoses (36%) being the most common. However [31], also concluded that erroneous diagnoses might actually represent comorbidity given that in 15% of cases, the initial diagnoses were maintained when the dementia diagnosis was established. The prolonged journey to obtaining an accurate diagnosis was variable and, for some, characterised by dismissal and poor communication among healthcare professionals. In a focus group study in the UK, both carers and people with dementia believed that getting a diagnosis depended on the healthcare professional who was consulted and geographic location [32]. Misdiagnosis had serious consequences for people with dementia and their carers, resulting in inappropriate referrals which was thought to waste time and restrict access to appropriate support [30, 31].

3.6 | Quality Ratings

All four qualitative papers included in this systematic review were assessed independently by two reviewers as being of high quality (scores of 80%+) using the CASP checklist (Table 2). However, there were several quantitative studies adjudged to be at risk of bias using the ROBINS-E dimensions tool. Nine of the 16 quantitative studies met three or fewer of the seven criteria described in the ROBINS-E dimensions tool (Table 3). Issues included, studies not using appropriate study design to answer the research questions, not selecting participant samples in a way to reflect the study population, or not taking missing data into consideration through sensitivity analysis etc. Although these risk of bias and quality assessments does not entirely negate the findings from these studies as drivers of potential change for services for people with dementia, it does have an impact on the strength of their findings and the potential generalisability of their conclusions. Furthermore, the assessment of several of the quantitative studies in this systematic review highlights the necessity to employ correct methods of selecting participants and sampling, using the appropriate study design to investigate the research question and understanding the need to be thorough in analyses and presenting robust, strong findings that have taken potential bias into account.

4 | Discussion

To our knowledge, this is the first systematic review on the topic of misdiagnosis in dementia. Collated evidence has highlighted several different issues affecting misdiagnosis of dementia, indicating misdiagnosis of dementia to be a serious issue for people living with dementia and their carers, as well as for healthcare professionals who are delivering diagnoses. However, this review has also highlighted many outstanding areas which require further attention to provide clearer clinical practice guidance to reduce misdiagnosis of dementia.

Research on misdiagnosis has only explored a small number of different dementia subtypes. To date, fronto-temporal dementia

TABLE 2 | Quality assessment of qualitative studies based on CASP.

	Q1	Q2	Q3	Q4	Q5	Q6	Q7	Q8	Q9	Q10	Score[a]
Hoppe [29]	Yes	Yes	Yes	Yes	Yes	Can't tell	Yes	Yes	Yes	Yes	90%
Novek and Menec [30]	Yes	Yes	Yes	Yes	Yes	Can't tell	Yes	Yes	Yes	Yes	90%
Sutcliffe et al. [32]	Yes	Yes	Yes	Yes	Yes	Can't tell	Yes	Yes	Yes	Yes	90%
van Vliet et al. [31]	Yes	Yes	Yes	Yes	Yes	Can't tell	Can't tell	Yes	Yes	Yes	80%

Note: Q1. Was there a clear statement of aims of the research? Q2. Is a qualitative methodology appropriate? Q3. Was the research design appropriate to address the aim of the research? Q4. Was the recruitment strategy appropriate to the aims of the research? Q5. Was the data collected in a way that addresses the research issue? Q6. Has the relationship between researcher and participants been adequately considered? Q7. Have ethical issues been taken into consideration? Q8. Was the data analysis sufficiently rigorous? Q9. Is there a clear statement of findings? Q10. How valuable is the research?

[a]"Yes" coded as 1; "no" and "can't tell" coded as 0

TABLE 3 | Quality assessment of quantitative studies based on ROBINS-E dimensions.

Author, year	Q1	Q2	Q3	Q4	Q5	Q6	Q7	Total[a]	% score
Aihara and Maeda [17]	No	No	Yes	No	Yes	Yes	Yes	3	42.9%
Barker et al. [26]	No	No	Yes	No	No	No	No	6	85.7%
Beber and Chaves [22]	No	No	No	No	Yes	No	No	6	85.7%
Besser and Galvin [18]	No	No	Yes	No	Yes	Yes	Yes	3	42.9%
Drabo et al. [24]	Yes	Yes	Yes	No	No	No	No	4	57.1%
Draper et al. [19]	No	No	No	No	Yes	Yes	No	5	71.4%
Hunter et al. [28]	Yes	Yes	No	Yes	Yes	Yes	Yes	1	14.3%
Lin et al. [10]	Yes	Yes	Yes	No	Yes	Yes	Yes	1	14.3%
Maserejian et al. [23]	Yes	No	Yes	No	No	Yes	Yes	3	42.9%
Mendez et al. [27]	Yes	Yes	Yes	No	Yes	No	No	3	42.9%
Sannemann et al. [25]	Yes	No	Yes	No	No	Yes	No	4	57.1%
Schrauf and Iris [20]	No	No	Yes	No	No	No	No	6	85.7%
Shinagawa et al. [21]	Yes	No	No	No	Yes	No	No	5	71.4%
Surendranathan et al. [12]	No	No	No	No	Yes	No	No	6	85.7%
Vergouw et al. [16]	No	No	No	No	No	No	No	7	100.0%
Zhu et al. [13]	Yes	Yes	Yes	No	Yes	Yes	Yes	1	14.3%

Note: Q1. Considering the study aim, was an appropriate study design used? Q2. Were participants selected in a way that study sample represents the study population? Q3. Did study report attrition rate? Q4. Was the potential effect of missing data taken into consideration (e.g., sensitivity analysis/other adjustment methods)? Q5. Was the main outcome assessed using reliable methods/assessment tools? Q6. Was an appropriate analysis strategy used to assess the relationship between predictors and outcomes? Q7. Were potential confounders taken into consideration in inferential analysis?

[a]Based on questions Q1 to Q7; "Yes" coded as 0; "no" and "unclear" coded as 1

has received the most attention [18, 22, 26, 27], followed by one study on Lewy Body dementia and PDD [12]. A Brazilian study reported that 90% of people with FTD and 80% with ADD experienced a misdiagnosis [22]. It is important to note that only nine and 10 people with FTD and ADD, respectively, were included by [22], which limits the generalisability of these findings. In addition, this study was conducted in Brazil, an upper-middle income country that experiences low resources into dementia and mental health, and notable difficulties in diagnosis and care [34, 35]. With limited research on individual subtypes, as well as overarching on rare dementia subtypes, more research is required to explore a wider range of subtypes perhaps by utilising existing health record data to explore misdiagnosis rates.

Whilst individual subtypes require further attention, early findings indicate particular challenges with misdiagnosis related to age for people suspected of YOD [18, 29–31]. This is linked to a lack of understanding in health care professionals, who may consider dementia to only be age-related and not occurring before the age of 65 (or 60 in lower- and middle-income countries). As result, people with an undiagnosed YOD may have to wait longer to receive a conclusive diagnosis, which can cause a great deal of distress [30]. Considering that general dementia care is often unsuitable for people with YOD [36–38], further awareness raising and relevant training has the potential to facilitate faster, more accurate, and more supportive experiences for people with YOD.

Whilst limited research has focussed on the economic impacts of misdiagnosis, two studies have shown that a misdiagnosis of dementia in the US was more cost-intensive in terms of health care usage than an initially correct subtype diagnosis [13, 28]. However, one study explored misdiagnosis based on Medicare data as opposed to a diagnosis by a health care professional [13]. Considering the substantial costs associated with dementia care alone, estimated at £13.5 billion annually in the UK [39] and $1313 billion globally (comprising medical, social care, and unpaid care costs; [1]), trying to reduce unnecessary costs associated with dementia is desirable. No single study emerged that had focussed on the economic impact of misdiagnosis on the individual and their unpaid carer. Whilst evidence exists on the financial pressures of caring for someone with dementia [40], a focus on misdiagnosis has to date not been explored specifically. Future research into misdiagnosis should investigate the economic costs and impacts.

Receiving any dementia diagnosis can be distressing [41], and requires both the person with the condition and their family to adjust and prepare. However, none of the studies on misdiagnosis have explored the emotional impacts of a misdiagnosis. Four studies have explored the experiences surrounding a diagnosis, highlighting particular challenges for those living with YOD and experiences of engaging with health care professionals. Considering the substantial emotional impact of dementia on both the person with dementia and the carer (i.e., [32]), it is surprising to not find any evidence on the emotional impacts of a misdiagnosis process and its long-term impacts on subsequent care. This is an urgent area for future research.

This systematic review benefits from two different researchers having screened each title and abstract, and full-text, having searched nine databases for relevant studies, and including evidence in four different languages where published. Final

searches were conducted in June 2023, which was within a year of submitting the manuscript. Only two studies had been published very recently, focussing on using poetry to describe the experiences of misdiagnosis [42] and correlates of missed or delayed diagnosis [43]. Creating 27 poems about experiences of misdiagnosis by people with rarer subtypes of dementia were found to be thought-provoking and helpful to over 90 surveyed health care professionals, to better understand the distress and lack of support received in the journey. This creative approach holds potential for workforce development and learning. Providing statistically significant evidence on the impact of personal characteristics on delayed diagnosis, Chen et al. [43] used Medicare data to show that people from a Black ethnic background and with higher levels of cognition at onset of dementia experienced higher odds of a delayed diagnosis. In contrast, those with a higher income or comorbidities experienced less likelihood of a delay. Findings from both studies, using different methodologies, feed into the evidence of inequalities in missed, delayed, and misdiagnosis of dementia, supporting evidence synthesised in this systematic review.

5 | Conclusions

Evidence is limited on misdiagnosis rates of different dementia subtypes and its personal and economic impacts. Evidence on economic and emotional impacts is minimal to non-existent, with economic impacts solely reported in the US. Findings from these studies may lack generalisability, that is the US Medicare system may not be relevant to all countries, highlighting a need for economic evaluations of misdiagnosis across other countries and healthcare contexts. However, existing evidence from 12 countries indicates that misdiagnosis is a frequent issue in rare dementias, and sometimes in ADD, with early evidence indicating substantial impacts on care costs, care access, and wellbeing. Diagnosing rarer dementias is important for people to receive the right care and support, albeit it can be difficult to do so for health care professionals. This is an urgent area of future investigation, and utilising EHR data combined with economic evaluations and qualitative explorations are key in developing guidance to reduce misdiagnosis and improve timely and correct subtype diagnosis.

Conflicts of Interest

The authors declare no conflicts of interest.

Data Availability Statement

Data sharing not applicable to this article as no datasets were generated or analysed during the current study.

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