

Cognitive Symptoms Preceding a Diagnosis of Early-Onset Frontotemporal Dementia: A Scoping Review

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Article

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Abstract

Background: Early-onset frontotemporal dementia (EO-FTD) presents before age 65 and is frequently misdiagnosed as psychiatric or behavioural disorders, delaying care.

Objective: This scoping review synthesizes research on EO-FTD's earliest cognitive symptoms from patients, companions (family and friends), healthcare professionals, and cognitive tests to promote early detection.

Methods: A systematic search of six databases (Medline, Embase, CINAHL, PsycInfo, Scopus, and Proquest Dissertations and Theses) identified 2197 studies of which 16 met inclusion criteria, encompassing 663 EO-FTD participants.

Findings: A total of 35 unique cognitive symptoms were identified. Memory, attention, and executive dysfunction were most frequently reported. Symptom terminology varied widely, often mirroring cognitive test phrasing, limiting clinical applicability. Many studies relied on cognitive test scores rather than detailed symptom descriptions, with patient and companion reports underrepresented.

Discussion: The findings underscore the need for standardized nomenclature, improved assessment tools, and greater inclusion of patient and companion perspectives to enhance EO-FTD early diagnosis and intervention strategies.

Résumé

Context: La démence frontotemporale précoce se manifeste avant l'âge de 65 ans et est souvent diagnostiquée à tort comme un trouble psychiatrique ou comportemental, ce qui retarde les soins.

Objectif: Cette revue de la portée synthétise la recherche sur les premiers symptômes cognitifs de l'EO-FTD chez les patients, les compagnons (famille et amis), les professionnels de la santé et les tests cognitifs afin de favoriser la détection précoce.

Méthodes: Une recherche systématique dans six bases de données (Medline, Embase, CINAHL, PsycInfo, Scopus et Proquest Dissertations and Theses) a permis d'identifier 2197 études, dont 16 répondaient aux critères d'inclusion, englobant 663 participants à l'EO-FTD.

Résultats: Au total, 35 symptômes cognitifs uniques ont été identifiés, la mémoire, l'attention et le dysfonctionnement exécutif étant les plus fréquemment signalés. Cependant, la terminologie des symptômes variait considérablement, reflétant souvent la formulation des tests cognitifs, ce qui limitait l'applicabilité clinique. De nombreuses études se sont appuyées sur des résultats de tests cognitifs plutôt que sur des descriptions détaillées des symptômes, les rapports des patients et des accompagnateurs étant sous-représentés.

Discussion: Les résultats soulignent la nécessité d'une nomenclature normalisée, d'outils d'évaluation améliorés et d'une plus grande inclusion des points de vue des patients et des accompagnateurs pour améliorer les stratégies de diagnostic et d'intervention précoces de l'EO-FTD.

Introduction

Early-onset dementia (EOD), defined as dementia with onset prior to the age of 65, is estimated to have affected 3.9 million people worldwide in 1990, rising to 7.8 million in 2021 (He et al., 2025). The increase is largely attributed to improvements in the diagnosis of EOD; however, it continues to be underdiagnosed (He et al., 2025; Leroy et al., 2021). EOD can have devastating consequences for families and create a high burden for caregivers, the healthcare system, and the overall economy (Antonioni et al., 2023; Hendriks et al., 2021). Within the spectrum of EOD,

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frontotemporal dementia (FTD) is the third most common form of dementia after Alzheimer's disease (AD) and dementia with Lewy Bodies (DLB) (Hendriks *et al.*, 2021). Typically manifesting between ages 45 and 65, and in some cases as early as the 20s, FTD usually spans a disease course of 8–10 years, although durations can range from 2 to 20 years (Antonioni *et al.*, 2023; Snowden *et al.*, 1996).

Timely diagnosis of FTD is critical yet challenging. Due to its involvement of the frontal lobes, FTD is generally characterized by changes in a person's personality, behaviour, and executive function as originally described in the Lund-Manchester Research Criteria (Neary, 1999; Snowden *et al.*, 1996). Contemporary frameworks, however, conceptualize FTD as a clinically heterogeneous spectrum with profiles that can include changes in language, social skills, motor skills, emotion processing, and other areas of cognitive impairment (e.g., semantic memory) (Bang *et al.*, 2015). Differentiating FTD from other conditions, such as depression, AD, or other psychiatric disorders, continues to be difficult (Hendriks *et al.*, 2021; Shelley *et al.*, 2007). Rosness *et al.* (2008) found that the interval from symptom onset to diagnosis in FTD ranged from 49.5 to 59.2 months on average, and Leroy *et al.* (2021) found the interval to be 37.8 months on average, with both studies reporting it was significantly longer than for AD. International research under the Genetic Frontotemporal Dementia Initiative (GENFI) has demonstrated that structural and cognitive changes can be detected up to 10 years before diagnosis (Rohrer *et al.*, 2015). GENFI studies also found that gene-specific cognitive profiles exist (Poos *et al.*, 2022) and that cognitive measures are sensitive indicators for tracking disease progression (Poos *et al.*, 2015). A more comprehensive understanding and tracking of the cognitive profile of early-onset FTD (EO-FTD), a major cause of EOD, has the potential to advance clinical practice by facilitating earlier diagnosis and intervention to ultimately enhance quality of life for people with EO-FTD.

FTD has been historically described as comprising three primary subtypes or variants encompassing FTD, each with distinct cognitive profiles. In the behavioural variant of FTD (bvFTD), cognitive impairments predominantly involve executive function, attention, abstraction, set shifting, response inhibition, planning and sequencing, verbal fluency, and decision-making. Semantic dementia (svFTD), or the semantic variant of primary progressive aphasia, primarily affects semantic memory, leading to difficulties with word comprehension, object recognition, face recognition, and sensory processing (e.g., tastes, smells, and tactile stimuli). Progressive non-fluent aphasia, or the non-fluent variant of primary progressive aphasia, is chiefly a language disorder characterized by speech apraxia with syntactic errors or word substitutions, though early deficits in calculation and verbal memory may also be evident, with executive dysfunction emerging later (Antonioni *et al.*, 2023; Snowden *et al.*, 1996).

Discussion persists in the categorization of FTD. For example, Snowden *et al.* (1996) noted the difficulty in including loss of insight as a criterion for bvFTD, although present in 85% of people with bvFTD, because of its evaluation reliability issues. They also noted the challenges with the interchangeable use of svFTD with the semantic variant of primary progressive aphasia, given that it is more of a semantic memory disorder than a primary aphasia. Similarly, Shelley *et al.* (2007) refer to bvFTD as the frontal variant of FTD (fvFTD) and further delineate fvFTD into apathetic and disinhibited types. More recently, additional less common subtypes have sometimes been included under the umbrella of FTD such as right temporal variant (rtvFTD), FTD associated with motor neuron disease (FTD-MND), corticobasal syndrome (CBS), and

progressive supranuclear palsy because of their frontal lobe involvement (Antonioni *et al.*, 2023). Within these expanded classifications, further clinical distinctions have emerged, with symptoms such as face recognition deficits now being more closely associated with rtvFTD rather than svFTD (Antonioni *et al.*, 2025; Ulugut *et al.*, 2024), highlighting the evolving nature of diagnostic frameworks. Such heterogeneity among the FTD variants further complicates the differentiation of FTD from other dementias or non-dementia-related aetiologies, hindering timely diagnosis and early intervention.

Cognitive symptoms remain a critical yet under-characterized component of EO-FTD. Although EO-FTD is increasingly recognized as involving a constellation of biomarkers and clinical symptoms (e.g., behavioural, motor) that extend beyond cognition, cognitive changes are often the first domain to be systematically assessed in clinical settings, are central to referral pathways for specialist evaluation, and are incorporated into most diagnostic testing at the time of presentation. Cognitive screening tests frequently used in dementia research and clinical practice, however, often do not adequately assess the executive dysfunction characteristic of FTD (Cavedoni *et al.*, 2020). As a result, early signs of FTD may be overlooked, especially in the absence of more comprehensive neuropsychological testing and detailed reports from patients and companions (family or friends). As cognitive symptoms are amenable to standardized measurement and longitudinal tracking, improved characterization of early cognitive presentation offers an opportunity to enhance early identification and inform the development of more sensitive assessment approaches within the broader multidimensional prodromal landscape of EO-FTD.

Objective and research questions

To clarify how cognition is currently characterized in EO-FTD, a scoping review was conducted to systematically map the presenting cognitive symptoms of EO-FTD prior to diagnosis. This review synthesizes findings from studies reporting cognitive symptoms as described by patients, their companions, health professionals, or derived from cognitive testing at the time of or prior to diagnosis, with the aim to identify gaps in current characterization that may limit early recognition, as well as key areas for further research and potential improvements in early diagnostic practices.

The original research questions were: (1) Which cognitive symptoms do adults, without other neurological conditions, experience prior to receiving a diagnosis of EOD? and (2) How are these cognitive symptoms identified – whether by the patient, companions (family or friends), a health professional, or via cognitive testing? Following title and abstract screening, due to the high volume of studies eligible for full-text review for all types of EOD, the study team refined the population in the first research question to focus on EO-FTD exclusively.

Methods

Design and eligibility criteria

This review followed the Preferred Reporting Items for Systematic Reviews and Meta-analysis extension for scoping reviews (Tricco *et al.*, 2018). The review protocol was not registered.

Cognitive symptoms were defined as 'observable or reported impairment in mental processes, such as memory, attention, language, executive function, and visuospatial abilities, which may

indicate underlying neuropsychological dysfunction' (Lezak et al., 2012). For the purposes of this scoping review, symptoms that are not inherently cognitive were excluded. Examples of these excluded symptoms include mental health or mood-related symptoms (e.g., anxiety, mood swings, sadness), specific language difficulties (e.g., aphasia), behavioural changes, psychiatric symptoms, and functional impairments (e.g., social dysfunction or difficulty with activities of daily living).

Studies were included in this scoping review if they met the following criteria: (1) reported specific cognitive symptoms experienced by individuals prior to, or at the time of, an EO-FTD diagnosis. Studies that only provided overall cognitive test scores or described cognitive symptoms solely after diagnosis or during disease progression were excluded; (2) were prospective studies reporting cognitive symptoms at initial presentation (prior to EO-FTD diagnosis) or retrospective studies detailing presenting cognitive symptoms at the time of EO-FTD diagnosis; (3) had a quantitative or qualitative study design. Case studies involving single patients, kindreds, or pedigrees were excluded; (4) identified cognitive symptoms using self-reports from patients, informant reports from companions (e.g., family or friends), cognitive test findings, or clinical observations; (5) were peer-reviewed journal articles or thesis dissertations written in English that involved adult human participants without other neurological conditions (e.g., comorbid brain injury, stroke, Down's Syndrome or Parkinson's, etc.). Articles focusing exclusively on brain imaging results, genetics, or non-cognitive symptoms (e.g., mood or neuropsychiatric features) of EOD were excluded.

Information sources and search strategy

Potentially relevant papers were identified through searches of the following online bibliographic databases: Medline, Embase, CINAHL, PsycInfo, Scopus, and Proquest Dissertations and Theses. The literature search was executed on November 28, 2024. The search strategies, drafted by an experienced research librarian (LD) and further refined through study team discussion, combined EOD and assessment/diagnosis with either cognition terms or qualitative research terms to be as sensitive as possible. No date, language, or study type limits were used. The full search strategy is included in the [Supplementary Materials](#). The final search results were exported into Covidence (Veritas Health Innovation, n.d.). Duplicates were removed automatically by Covidence or manually by the reviewers.

Screening and eligibility

Two reviewers (TM and FA) independently applied the predefined inclusion and exclusion criteria to an initial set of 40 abstracts in parallel to calibrate their understanding and ensure consistent application of the eligibility criteria. The reviewers then proceeded with independent title and abstract evaluations of all remaining records using the same a priori criteria. Publications were advanced to full-text review if they met the refined inclusion criteria – specifically, focusing on EO-FTD and excluding case studies involving single patients, kindreds, or pedigrees. Both reviewers then assessed the full texts to confirm eligibility for data extraction, and data were extracted by both reviewers. Any disagreements during study selection or data extraction were resolved through consensus and, when necessary, discussion with the broader study team.

Data-charting process

A data-charting form was collaboratively developed by the two reviewers in consultation with the study team to systematically extract information on cognitive symptoms reported prior to EO-FTD diagnosis, as well as the sources from which these symptoms were identified.

For each publication, the form also captured the study characteristics such as year of publication, study objective, study type and design, method of analysis, study setting, country, number of EO-FTD participants, as well as EO-FTD participant demographic characteristics including sex, education, ethnicity (when reported), and age at onset or first clinical presentation. The two reviewers independently and iteratively reviewed the texts, charted the data, and subsequently updated the form based on their findings, ensuring consistency and accuracy throughout the process.

Data analysis and synthesis

Comprehensive descriptive summaries were generated for each included study to support synthesis and interpretation. A formal appraisal of the methodological quality or risk of bias of included studies was not conducted given the objective of this scoping review to map and summarize the existing body of knowledge regarding the presenting cognitive symptoms of EO-FTD. This follows established methodological guidance for scoping reviews on clinical topics (Tricco et al., 2016).

Cognitive symptoms experienced by adults prior to an EO-FTD diagnosis were extracted from the included studies. All cognitive symptoms are reported as originally named in each of the studies. The frequency with which each cognitive symptom was measured, along with the frequency of each source type. In addition, a breakdown of the studies by country is provided.

Results

Selection of sources of evidence

Six databases (Scopus, Embase, MEDLINE, PsycInfo, CINAHL, and Proquest Dissertations and Theses) were searched originally for EOD (all types) and yielded 2,197 studies after duplicate removal. Following title and abstract screening by the two reviewers, 313 articles were identified as eligible for full-text review across all EOD types, including case studies. Given the study team's decision to exclusively focus on EO-FTD, these 313 studies were imported into a separate Covidence instance. Title and abstract screening were then repeated with the additional inclusion criteria of FTD and the exclusion criteria of case studies applied. Ultimately, 34 studies proceeded to full-text review, of which 16 studies met the eligibility criteria for inclusion. Studies were excluded at full-text review for the following reasons: not written in English ($n = 2$), failure to identify cognitive symptoms ($n = 13$), inability to distinguish EO-FTD symptoms from late-onset FTD symptoms ($n = 2$), or failure to differentiate the presenting cognitive symptoms for EO-FTD from other dementia types ($n = 1$). [Figure 1](#) presents the PRISMA flow diagram outlining the study selection process.

Study characteristics

Characteristics of each study were identified, including publication date, study objective, study design, study setting and country, and

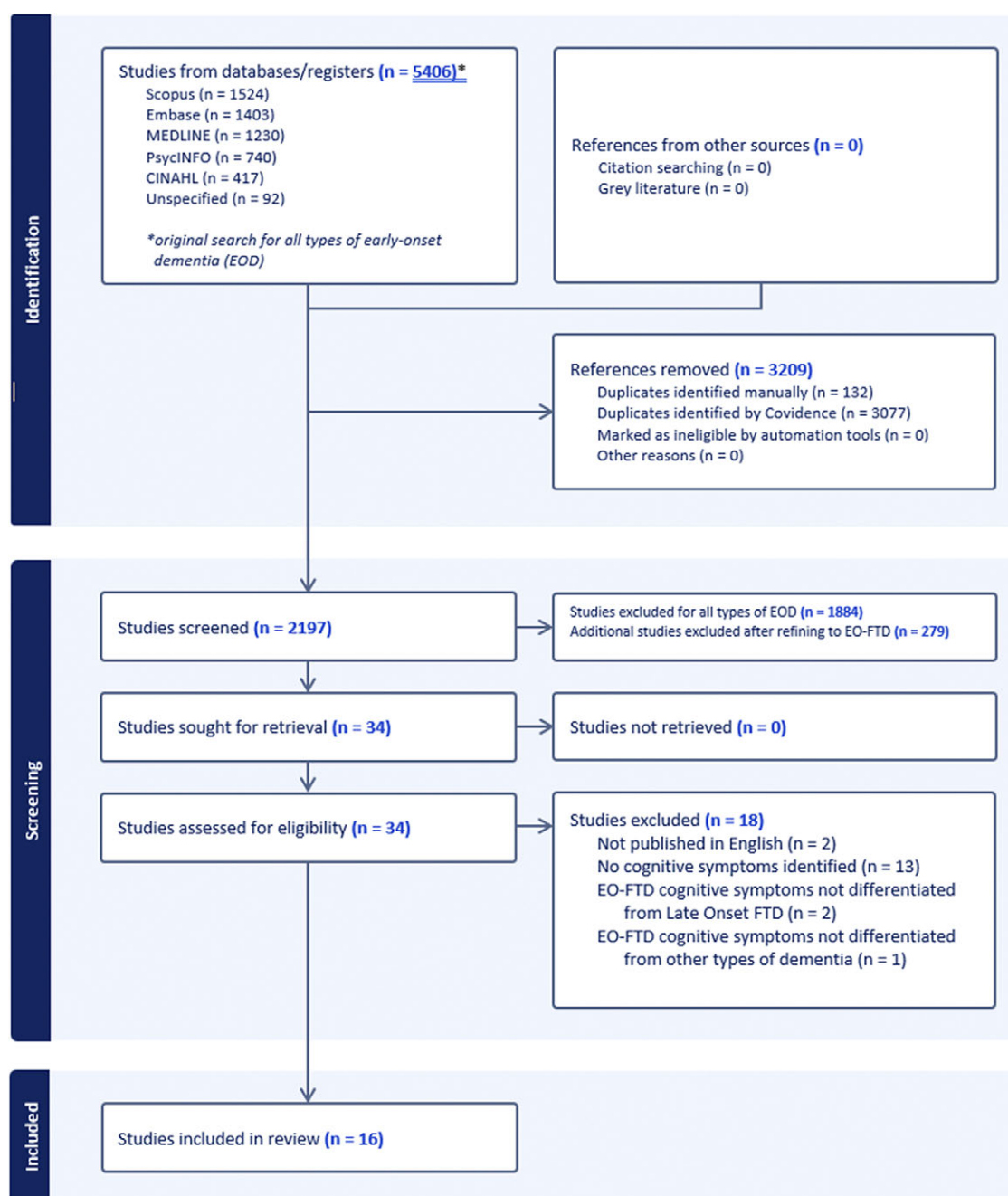


Figure 1. PRISMA flow diagram of study selection process.

method of analysis (Table 1). The following is a synthesis of these findings.

Publication dates ranged from 2007 to 2024, with half of the studies ($n = 8$) published between 2007 and 2012 (Chow et al., 2011; Hornberger et al., 2009; Panegyres & Frencham, 2007; Panegyres et al., 2007; Rosness et al., 2008; Shelley et al., 2007; Shinagawa et al., 2008; Snowden et al., 2011). Research settings were internationally distributed (Figure 2).

Clinical profiles of dementia were examined in the majority of studies, with 11 of the 16 studies explicit in this aim (Edahiro et al., 2023; Heikkinen et al., 2024; Panegyres, 2021; Panegyres & Frencham, 2007; Panegyres et al., 2007; Rivas Nieto, 2014; Serrano et al., 2014; Shelley et al., 2007; Shinagawa et al.,

2008; Snowden et al., 2011; van Gils et al., 2023). Six of these specifically stated an intent to identify cognitive or neuropsychological symptoms as part of the clinical profile (Heikkinen et al., 2024; Panegyres, 2021; Rivas Nieto, 2014; Serrano et al., 2014; Shelley et al., 2007; Shinagawa et al., 2008).

Study designs were observational, descriptive cross-sectional, and qualitative in nature. The majority of studies employed observational designs ($n = 13$) (Falgas et al., 2019; Heikkinen et al., 2024; Hornberger et al., 2009; Panegyres, 2021; Panegyres & Frencham, 2007; Panegyres et al., 2007; Rivas Nieto, 2014; Rosness et al., 2008; Serrano et al., 2014; Shelley et al., 2007; Shinagawa et al., 2008; Snowden et al., 2011; van Gils et al., 2023), including prospective ($n = 6$) (Falgas et al., 2019; Panegyres, 2021; Panegyres &

Table 1. Overview of included studies

Author(s) (year)	Study title	Study objective	Study design	Study setting country	Method of analysis
Chow et al. (2011)	An international needs assessment of caregivers for frontotemporal dementia	To guide the development of public awareness and caregiver support resources for FTD syndromes	Descriptive: retrospective cross-sectional survey	Online Canada, USA, UK, and Europe	Descriptive statistics
Edahiro et al. (2023)	Initial symptoms of early-onset dementia in Japan: nationwide survey	To investigate initial pre-diagnostic symptoms of EOD for each dementia subtype as a part of the JNEP study	Descriptive: retrospective cross-sectional survey	Online Japan	Descriptive statistics, chi-square tests for associations between types, additional multivariate analyses
Falgas et al. (2019)	Clinical applicability of diagnostic biomarkers in early-onset cognitive impairment	To describe the diagnostic implications of using diagnostic biomarkers on clinical practice in early cognitive impairment to determine their added diagnostic confidence and the patient's tolerance of the procedures	Observational: prospective cohort study	Alzheimer's Disease & Other Cognitive Disorders Unit at the Hospital Clinic Barcelona Spain	Descriptive statistics, comparison with t-tests, chi-square tests for associations
Heikkinen et al. (2024)	Neuropsychological profiles, aetiologies, and medical comorbidities in early-onset dementia and cognitive impairment: a memory outpatient clinic cohort study	To investigate the neuropsychological profiles, aetiologies, and comorbidities of an unselected cohort of memory clinic patients (≤ 65 years at symptom onset)	Observational: prospective and retrospective cohort study	Memory outpatient clinics in Oulu and Kuopio University Hospitals Finland	Descriptive statistics, associations, comparison with Pearson chi-square, nonparametric tests
Hornberger et al. (2009)	Can progressive and non-progressive behavioural variant frontotemporal dementia be distinguished at presentation?	To investigate whether the two groups (progressors and non-progressors) of bvFTD can be discriminated by their clinical features at first presentation and caregiver info from Cambridge Behavioural Inventory	Observational: retrospective cohort comparison study	Archival clinical data of the Early-Onset Dementia Clinic Cambridge United Kingdom	Descriptive statistics, t-tests for parametric variables, chi-square, and Mann–Whitney tests for nonparametric categorical data
Johannessen et al. (2017)	Experiences and needs of spouses of persons with young-onset frontotemporal lobe dementia during the progression of the disease	To examine the partners of people with FTLD experiences and needs for assistance in daily life	Retrospective Qualitative interviews	Seven memory clinics, one municipality dementia team, & a nursing home for YOD Norway	Qualitative analysis for themes
Panegyres (2021)	The clinical spectrum of young onset dementia points to its stochastic origins	To define the clinical diversity of young onset dementia in terms of its demography, range of presentations, neurological examination findings, comorbidities, medical history, cognitive findings, imaging abnormalities both structural and functional, electroencephalographic (EEG) data, neuropathology, and genetics	Observational: prospective cohort study	Specialist community-based clinics established by the author with the University of Western Australia Australia	Descriptive statistics, chi-square tests for associations, and t-tests for comparison
Panegyres and Frencham (2007)	Course and causes of suspected dementia in young adults: a longitudinal study	To analyse a cohort of young patients assessed consecutively with the question of dementia	Observational: prospective cohort study	Early-onset dementia multidisciplinary clinic at the Neurosciences Unit in Perth Australia	Descriptive statistics
Panegyres et al. (2007)	The clinical differentiation of frontotemporal dementia from psychiatric disease	To develop clinical profiles to distinguish between FTD and psychiatric disorders, particularly when frontal and/or temporal lobar atrophy is observed on neuroimaging	Observational: retrospective case control comparison study	Early-onset dementia multidisciplinary clinic at the Neurosciences Unit in Perth Australia	Descriptive statistics, trend analysis

(Continued)

Table 1. Continued

Author(s) (year)	Study title	Study objective	Study design	Study setting country	Method of analysis
Rivas Nieto (2014)	Frontotemporal dementia: clinical, neuropsychological, and neuroimaging description	To describe the clinical, neuropsychological, and imaging findings from a group of patients diagnosed with FTD	Observational: retrospective cohort study	Psychiatric Hospital Universitario del Valle in Cali Colombia	Descriptive statistics, chi-square tests for associations
Rosness et al. (2008)	Frontotemporal dementia—a clinically complex diagnosis	To examine the time taken to establish a clinical diagnosis FTD compared with a clinical diagnosis of early-onset Alzheimer's Disease (AD), and to explore factors that can explain why it takes a long time to diagnose FTD	Observational: retrospective cohort study	Four memory clinics and two neurology departments Sweden & Norway	Descriptive statistics, chi-square tests for associations, t-tests, or nonparametric tests for comparisons
Serrano et al. (2014)	Onset age and clinical heterogeneity of dementias: a diagnostic and therapeutic approach	To evaluate the clinical characteristics of patients with senile dementia onset (> 65 years) and presenile onset (<65 years) and to compare their neuropsychiatric and neuropsychological profiles according to onset age	Observational: prospective and retrospective cohort comparison study	Research Laboratory of Memory Argentina	Chi-square for associations, descriptive statistics, t-tests for comparisons, Mann–Whitney U test when normality assumptions not met
Shelley et al. (2007)	Frontotemporal dementia in Oman: cognitive behavioural profile and neuroimaging characteristics; a prospective hospital-based study	To investigate the cognitive and behavioural profiles of patients diagnosed with the frontal variant of FTD in Oman. To examine the neuroimaging characteristics (using MRI and SPECT) associated with this condition in these patients	Observational: prospective cohort study	Cognitive Behavioural Clinic Oman	Descriptive statistics
Shinagawa et al. (2008)	Cognitive function and psychiatric symptoms in early- and late-onset frontotemporal dementia	To compare cognitive function and psychiatric symptoms in consecutive patients with early- and late-onset FTD attending a memory clinic in Japan	Observational: prospective cohort comparison study	Higher Brain Function Clinic of the Department of Neuropsychiatry, Ehime University Hospital Japan	Descriptive statistics, t-tests, and Mann–Whitney U test for comparisons, chi-square tests for associations
Snowden et al. (2011)	The clinical diagnosis of early-onset dementias: diagnostic accuracy and clinicopathological relationships	To outline the principles that guided differential diagnosis in patients attending a specialist early-onset dementia clinic	Observational: retrospective cohort study	Neuroscience Centre at a Manchester University teaching hospital United Kingdom	Descriptive statistics
Van Gils et al. (2023)	Young-onset dementia in memory clinics in the Netherlands: study design and description of PRECODE-GP	To leverage the Dutch memory clinic infrastructure to set up a national database of patients with YOD, to provide accurate data on the occurrence and characteristics of YOD and provide a basis for future in-depth studies on YOD	Observational: prospective cohort study	39 memory clinics Netherlands	Descriptive statistics, group differences with ANOVA or chi-square tests as appropriate, linear regression analysis for MMSE scores

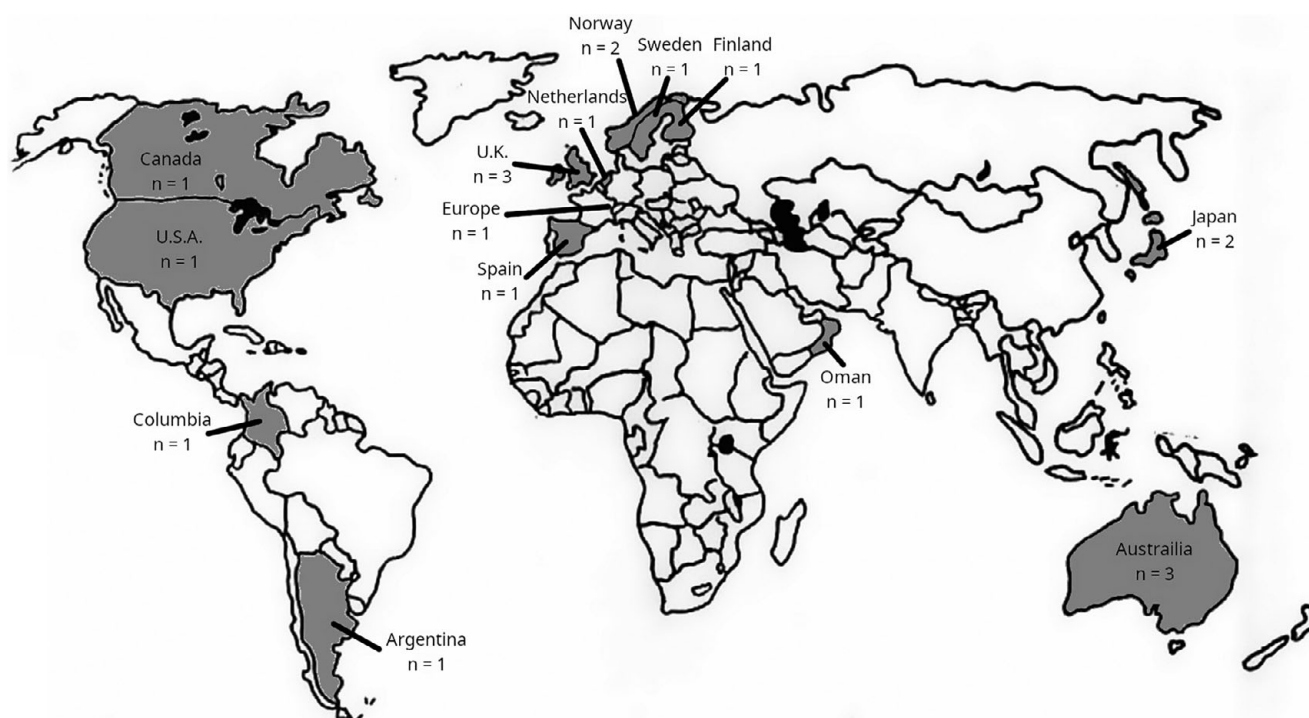


Figure 2. Distribution of study settings by country.

Frencham, 2007; Shelley et al., 2007; Shinagawa et al., 2008; van Gils et al., 2023), retrospective ($n = 4$) (Hornberger et al., 2009; Rivas Nieto, 2014; Rosness et al., 2008; Snowden et al., 2011), prospective and retrospective ($n = 2$) (Heikkinen et al., 2024; Serrano et al., 2014), and one case control study (Panegyres et al., 2007). Two studies used descriptive, cross-sectional survey designs (Chow et al., 2011; Eda Hiro et al., 2023) and one study employed a qualitative interview approach (Johannessen et al., 2017). Nearly, all studies ($n = 15$) reported analyses of cognitive symptoms using descriptive statistics, apart from the qualitative study by Johannessen et al. (2017).

Cognitive symptoms preceding EO-FTD diagnosis

A total of 35 uniquely named cognitive symptoms were identified as presenting symptoms of EO-FTD. The most frequently reported cognitive impairments were in memory (all studies), followed by attention ($n = 10$) (Heikkinen et al., 2024; Hornberger et al., 2009; Johannessen et al., 2017; Panegyres et al., 2007; Rivas Nieto, 2014; Rosness et al., 2008; Serrano et al., 2014; Shelley et al., 2007; Snowden et al., 2011; van Gils et al., 2023) and then executive function ($n = 7$) (Falgas et al., 2019; Heikkinen et al., 2024; Panegyres et al., 2007; Rivas Nieto, 2014; Serrano et al., 2014; Shelley et al., 2007; van Gils et al., 2023). All other cognitive symptoms were mentioned in only one to three studies.

The cognitive symptoms identified in each study and their reported sources are summarized in Table 2, along with the EO-FTD participant demographics (number, sex, and education, when reported). Ethnicity was not included in Table 2 as it was only reported in one study (van Gils et al., 2023) with 77% of the participants being Caucasian. Across the 16 included studies, there were 663 EO-FTD participants, with half of the studies ($n = 8$) (Falgas et al., 2019; Heikkinen et al., 2024; Johannessen et al., 2017; Panegyres et al., 2007; Rivas Nieto, 2014; Serrano et al., 2014;

Shelley et al., 2007; Shinagawa et al., 2008) comprising 25 or fewer participants.

Sources of presenting cognitive symptoms

Cognitive symptom data were variably derived. Ten studies (Falgas et al., 2019; Heikkinen et al., 2024; Panegyres & Frencham, 2007; Panegyres et al., 2007; Rivas Nieto, 2014; Serrano et al., 2014; Shelley et al., 2007; Shinagawa et al., 2008; Snowden et al., 2011; van Gils et al., 2023) obtained data from cognitive tests. Three additional studies (Hornberger et al., 2009; Panegyres, 2021; Rosness et al., 2008) performed cognitive testing but reported overall scores only, with no detail on the specific cognitive domains affected.

Cognitive symptom identification relied on various sources across studies. Figure 3 illustrates the frequency and proportion of studies that utilized each source. Notably, nine of the 16 studies (Chow et al., 2011; Falgas et al., 2019; Hornberger et al., 2009; Johannessen et al., 2017; Panegyres & Frencham, 2007; Rivas Nieto, 2014; Serrano et al., 2014; Shelley et al., 2007; Shinagawa et al., 2008) relied on a single source for symptom identification – six of these studies (Falgas et al., 2019; Panegyres & Frencham, 2007; Rivas Nieto, 2014; Serrano et al., 2014; Shelley et al., 2007; Shinagawa et al., 2008) exclusively reported cognitive test results and three studies relied solely on companion reports (Chow et al., 2011; Hornberger et al., 2009; Johannessen et al., 2017). Patient report was included in only six of the 16 studies (Eda Hiro et al., 2023; Panegyres, 2021; Panegyres et al., 2007; Rosness et al., 2008; Snowden et al., 2011; van Gils et al., 2023).

Dementia screening instruments were used in many studies. Among the 10 studies reporting cognitive symptoms from cognitive tests, six studies (Falgas et al., 2019; Panegyres & Frencham, 2007; Rivas Nieto, 2014; Shelley et al., 2007; Shinagawa et al., 2008; van Gils et al., 2023) utilized traditional dementia screening instruments (e.g., MMSE, MoCA, or Clock Drawing Test), with two of

Table 2. Cognitive symptoms preceding EO-FTD diagnosis and source of symptoms

Author(s) (year)	EO-FTD study participants: number (<i>n</i>) sex education (mean years (yrs))	EO-FTD participant mean age of onset or first presentation in years (SD or range)	Cognitive symptoms preceding diagnosis	Source of symptoms
Chow et al. (2011)	<i>n</i> = 79 26 male 37 female 16 skipped question Education not stated	58 (range 28–78)	Thinking & judgement, Memory	Companion report
Eda Hiro et al. (2023)	<i>n</i> = 83 Sex not stated Education not stated	57.2 (5.6)	Memory, Word generation	Patient report, Companion report
Falgas et al. (2019)	<i>n</i> = 5 20% female Education not stated	56.4 (3.7)	Executive function	Cognitive test(s): MMSE, Neuropsychological testing
Heikkinen et al. (2024)	<i>n</i> = 10 Sex not stated for FTD group Education not stated	Onset age not stated for FTD group	Verbal immediate memory, Verbal delayed memory, Visual immediate memory, Visual delayed memory, Working memory, Attention, Processing Speed, Reasoning, Executive function	Clinician observations, Cognitive test(s): Neuropsychological testing
Hornberger et al. (2009)	<i>n</i> = 71 52 male 19 female Education not stated	Progressors: 57.6 (8.5) Non-progressors: 54.1 (7.6)	Memory, Attention-orientation	Companion report *cognitive tests performed but only overall scores reported in study
Johannessen et al. (2017)	<i>n</i> = 16 10 male 6 female Education not stated	Mean not stated (range 47–65)	Attention, Route-finding, Memory	Companion report
Panegyres (2021)	<i>n</i> = 58 (all bvFTD) 39 male 19 female Education: <10 yrs, <i>n</i> = 21 yrs 11–12 yrs, <i>n</i> = 16 yrs >=13 yrs, <i>n</i> = 21 yrs	57.1 (5.66)	Memory	Patient report, Companion report, Cognitive test(s) *cognitive tests performed but only overall scores presented, with exception of memory discussed
Panegyres and Frencham (2007)	<i>n</i> = 43 21 male 22 female Education not stated	58.7 (range 31.52–61.4)	Memory, Short-term memory loss, Word generation	Cognitive test(s): MMSE, Neuropsychological testing, Wechsler Adult Intelligence Scale – Revised
Panegyres et al. (2007)	<i>n</i> = 13 7 male 6 female Education not stated	Not stated	Attention, Concentration, Memory, Executive function, Verbal learning, Verbal reasoning, Memory autobiographic, Memory semantic	Patient report, Companion report, Clinician observations, Cognitive test(s): Rey Complex Figure Test Rey Auditory Verbal Learning Test Controlled Oral Word Association Test Wechsler Adult Intelligence Scale – III
Rivas Nieto (2014)	<i>n</i> = 21 11 male 10 female Education not stated	59.8 (2.7 yrs mean of symptoms prior to diagnosis)	Memory, Attention, Executive function, Calculation	Cognitive test(s): MMSE, Clock Drawing Executive Test (CLOX)

(Continued)

Table 2. Continued

Author(s) (year)	EO-FTD study participants: number (n) sex education (mean years (yrs))	EO-FTD participant mean age of onset or first presentation in years (SD or range)	Cognitive symptoms preceding diagnosis	Source of symptoms
Rosness et al. (2008)	Norway: n = 32 17 male 15 female Education: 12.1 yrs (2.8) Sweden: n = 20 8 male 12 female Education not stated	Norway: 59.1 (4.7) Sweden: Not stated	Memory, Attention	Patient report, Companion report *cognitive testing was performed but only overall score was reported
Serrano et al. (2014)	n = 21 Sex not stated Education: 10.76 yrs	58.52 (not stated)	Memory, Verbal fluency, Verbal reasoning, Executive function, Attention	Cognitive test(s): Verbal IQ, Full test IQ
Shelley et al. (2007)	n = 6 (all bvFTD) 3 male 3 female All with elementary school education	51.5 (range 47–63)	Verbal fluency, Attention-orientation, Memory, Executive function, Verbal learning, Word generation, Abstract to categories	Cognitive test(s): Clinical Dementia Rating Scale MMSE Addenbrooke's Cognitive Examination Frontal Assessment Battery Clock Drawing Test Stroop's Colour Word Sorting Test Reitan's Trail Making Test A/B *some cognitive tests performed only reported the overall score
Shinagawa et al. (2008)	n = 21 11 male 10 female Education: 11 yrs	63 (8.3)	Verbal delayed memory	Cognitive test(s): MMSE Clinical Dementia Rating RCPM Digital span tests ADAS-Jcog Clock Drawing Test Word Fluency Test *only MMSE and RCPM scores reported in study results; overall score only for RCPM
Snowden et al. (2011)	n = 96 Sex not specific to EO-FTD Education not stated	57 (9)	Memory, Verbal immediate memory, Verbal delayed memory, Attention, Comprehension, Judgement, Cognitive endurance, Shifting attention, Sequencing, Verbal memory, Recognition, Implementation and reflection, Poor organization, Concreteness, Impulsivity, Verbal fluency, Free recall, Perseveration	Patient report, Clinician observations, Cognitive test(s): Manchester Neuropsychological Profile
Van Gils et al. (2023)	n = 87 (20 with bvFTD) 44 male 43 female Education 11 yrs (3.1)	61 (6)	Memory, Executive, Attention, Concentration	Patient report, Cognitive test(s): MMSE MoCA

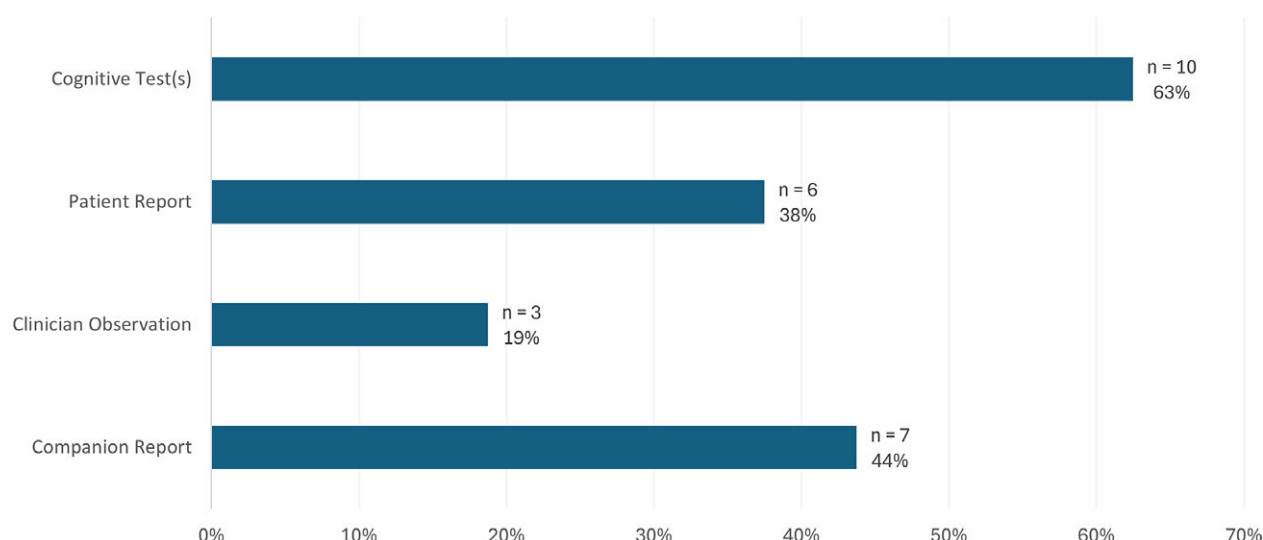


Figure 3. Frequency and proportion of studies utilizing each source of cognitive symptom data (n = 16).

these studies (Rivas Nieto, 2014; van Gils *et al.*, 2023) depending solely on dementia screening tests.

Summary

Sixteen internationally distributed studies were included in this scoping review when mapping the literature on the presenting cognitive symptoms of EO-FTD, spanning publication dates from 2007 to 2024. Few studies had the specific aim to assess the cognitive symptoms of EO-FTD. In response to this scoping review's first research question, 35 uniquely named cognitive symptoms were found to be experienced by adults, without other neurological conditions, prior to receiving a diagnosis of EO-FTD. Memory, attention, and executive function were the most frequently reported. In response to the second research question on how cognitive symptoms were identified, it was found that the majority of studies relied on a single source of data, with patient reports notably underrepresented. Cognitive tests provided symptom data in the majority of studies, and dementia cognitive screening tests were often utilized.

Discussion

There is a notable paucity of studies that report the specific cognitive symptoms preceding a diagnosis of EO-FTD with only 16 studies included in this scoping review. Studies were internationally dispersed enhancing generalizability but highlighted a need for more in-depth, country-specific research that explicitly aims to identify the cognitive symptoms preceding EO-FTD.

Nomenclature to describe presenting cognitive symptoms was inconsistent across studies. The wording of the 35 cognitive symptoms that were found to present prior to EO-FTD diagnosis frequently mirrored the terms used in the cognitive tests employed, resulting in a variety of labels for what appear to be similar cognitive constructs (e.g., verbal fluency with word generation; reasoning with verbal reasoning; attention with concentration). This inconsistency underscores the need for harmonization of cognitive symptom terminology across research, particularly for cross-study comparisons and clinical translation.

Broad cognitive domains such as 'memory' or 'attention' were reported as impaired in most studies rather than delineating more

precise symptoms (e.g., verbal delayed memory, working memory, or prospective memory under memory; sustained attention, selective attention, or divided attention under attention). This lack of specificity is clinically important, as these more specific cognitive impairments are likely to affect daily functioning in different ways. The ambiguity that is inherent in the reporting of broad cognitive domains is also particularly problematic when reported by patients or companions because these terms can encompass a wide range of distinct cognitive functions (Rasmussen *et al.*, 2019). Patients and companions typically lack clinical vocabulary and rely on general language to describe changes they observe. They are unlikely to use specific terms like 'working memory' or 'selective attention', which creates a communication gap between lived experience and clinical interpretation. This mismatch in language complicates a clinician's ability to accurately interpret reported symptoms, especially when trying to distinguish between neurodegenerative conditions with overlapping presentations (Rasmussen *et al.*, 2019).

These communication challenges are supported in previous research. Rasmussen *et al.* (2019) found through interviews with family caregivers of individuals with FTD that they often described early symptoms in vague or non-clinical terms, contributing to diagnostic delays. Similarly, Forbush *et al.* (2013) demonstrated how patients' own words are frequently transformed or lost when translated into clinical terminology in documentation. Both studies underscore the need to develop a shared framework for symptom reporting that can bridge clinical and lay language. Such a framework might link functional examples of cognitive changes to the specific cognitive symptom (e.g., 'difficulty following multi-step directions' to illustrate working memory issues or 'trouble focusing in conversations' to represent sustained attention difficulties) fostering a shared understanding between patients, companions, and clinicians.

Variability in the cognitive assessments employed across studies was found in this scoping review, even when targeting the same cognitive domain. For instance, one study may assess verbal delayed memory through a word list recall (e.g., Rey Auditory Learning Test in Panegyres *et al.*, 2007), while another uses story recall (e.g., The Wechsler Memory scale as part of neuropsychological testing in Heikkinen *et al.*, 2024); both target memory but in functionally different ways. In addition, the use of cognitive screening tests in studies, while quick to administer, inherently restricts

the number of cognitive symptoms that can be identified. This can contribute to potentially missing presenting cognitive symptoms of EO-FTD that are either not assessed or inadequately assessed by these screening tests (e.g., executive function, a common symptom of EO-FTD) (Cavedoni et al., 2020). Even in studies where more detailed neuropsychological testing was reported to have been performed, the specific tests administered were not consistently reported. In addition, while overall scores were often reported in studies as they facilitated group comparisons, aggregate scores lack the granularity needed for a more nuanced clinical understanding of presenting cognitive symptoms. In contrast, the Snowden et al. (2011) study which employed the Manchester Neuropsychological Profile, a bespoke assessment instrument designed to characterize different forms of dementia, offered the most detailed list of initial cognitive symptoms, underscoring the potential value in employing comprehensive cognitive assessment tools in research and clinical practice.

The source of cognitive symptoms within a study also often lacked diversity with the majority of studies relying on a single source of cognitive symptoms data, often cognitive tests. The lack of inclusion of first-person perspectives and the lack of more qualitative studies can limit the understanding of how cognitive symptoms are experienced and described outside of clinical settings – an important consideration for both clinical diagnosis and the ability to develop sensitive AI-based detection tools from clinical documentation review that could augment the clinical assessment process in future.

The lack of consistency in cognitive nomenclature, test selection, and sources across studies limits comparability and weakens the foundation for synthesizing symptom profiles that could aid in early detection or differential diagnosis of EO-FTD. Collectively, these issues expose key gaps in the current approach to characterizing cognitive symptoms that precede EO-FTD diagnosis. They highlight the need for greater standardization in both the terminology used to describe cognitive symptoms and the assessment tools and sources employed across studies.

Limitations

Several potential limitations were identified. First, only articles published in English were included in the full-text review, resulting in the exclusion of two potentially relevant studies published in French and Spanish (Croisile et al., 2012; Vilalta-Franch et al., 2008). Consequently, potentially relevant research could have been omitted, introducing a language bias that may have influenced the comprehensiveness of our findings.

Second, sample sizes varied considerably across the included studies, with half reporting fewer than 25 participants. The limited sample sizes reduce the generalizability of findings and can increase the risk of over-representing or missing certain cognitive symptoms due to small study populations. Furthermore, some of the included studies were not primarily designed to characterize the presenting cognitive symptoms of EO-FTD with FTD often a subset of a broader study including more dementia types, highlighting a gap in the literature regarding the description of cognitive symptoms that precedes diagnosis of EO-FTD on an international level.

Third, the lack of differentiation in the studies by the FTD variants when reporting the presenting cognitive symptoms is another limitation. Given bvFTD, svPPA, and nfvPPA exhibit distinct cognitive profiles, the absence of subtype-specific symptom reporting may obscure meaningful clinical distinctions.

Future directions and recommendations

More standardized, granular investigations into the presenting cognitive symptoms in EO-FTD are identified as needed from this scoping review. Future studies should aim for increased sample sizes, greater language inclusivity, and clearer differentiation by FTD subtype. More detailed and transparent reporting of cognitive testing results is needed to support synthesis across studies and advance early identification and differential diagnosis of EO-FTD.

Incorporating diverse sources of symptom information such as patient report, companion report, and clinician observations in addition to cognitive tests will enhance the accuracy and richness of symptom characterization. Within studies, cognitive symptoms need to be reported more precisely than broad cognitive domains such as ‘memory’ or ‘attention’. Standardizing the use of both cognitive symptom terminology and assessment tools across research and clinical practice would further enhance and support earlier and more reliable identification of EO-FTD.

Future scoping reviews by the study team will address the presenting cognitive symptoms of other types of EOD and include broader comparisons across dementia types. Case studies will also be reviewed as their exclusion may have limited the identification of rich, detailed cognitive symptom descriptions often captured in individual case reports, particularly those based on direct observations or firsthand accounts from patients and their companions.

For clinical practice, the development of standardized and accessible cognitive profiles tailored to FTD subtypes as well as different types of dementia to aid in differential diagnosis should be prioritized. These profiles should incorporate shared nomenclature that aligns with clinical frameworks and can be used across patients, companions, and clinicians to promote clearer communication and improve early recognition. The development of supportive tools, such as visual aids, and patient-friendly symptom lists with corresponding functional activity impacts can help patients and companions more precisely articulate cognitive changes. Clinical guidelines should recommend the use of specific standardized cognitive tests appropriate to the suspected dementia types and subtypes.

Future early detection strategies will likely require integration of cognitive symptom profiles with scalable, non-invasive diagnostic tools given the current non-specificity of early cognitive symptoms in EO-FTD. Emerging approaches such as blood-based biomarkers (Antonioni et al., 2025), brain stimulation (Benussi et al., 2021), and other minimally invasive techniques that have shown promise for population-level screening may help refine referral pathways when combined with sensitive cognitive characterization. Moreover, this underscores the importance of developing well-defined cognitive phenotypes that can be meaningfully integrated into multimodal diagnostic frameworks.

Conclusions

This scoping review reveals several critical insights and persistent gaps in the current literature on the presenting cognitive symptoms of EO-FTD. Although 35 distinct cognitive symptoms were identified, the terminology used to describe these symptoms varied across studies often resulting in symptoms that appeared to overlap in cognitive constructs. Most studies did not precisely delineate the impaired cognitive domains, and symptom data were most often from a single source, typically cognitive tests, both limiting the depth and contextual understanding of EO-FTD. Overall, these insights point to the need for the development of a unified, standardized framework for cognitive symptom nomenclature to foster a shared understanding among patients, companions, and

clinicians. Greater standardization and consistency in cognitive test selection are needed, as well as the integration of more comprehensive, methodologically diverse approaches, including qualitative methods. Addressing these gaps to better capture and standardize the complex presentation of the cognitive symptoms that precede the diagnosis of EO-FTD can assist to improve timely and accurate diagnosis.

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