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Receiving a diagnosis of young onset dementia: a scoping review of lived experiences

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Receiving a diagnosis of young onset dementia: a scoping review of lived experiences

Objectives: The personal experience of receiving a diagnosis of young onset dementia (YOD) is often overlooked in a complex assessment process which can require substantial investigation. To understand the lived experiences of younger people undergoing assessment and receiving a diagnosis of dementia, we undertook a thematic synthesis of qualitative studies published until November 2018. The aim of this review was to inform a Delphi study with younger people diagnosed with dementia to learn how diagnostic process could be improved, and to identify the strengths and weaknesses of current approaches and help educate professionals concerning key issues.

Method: Systematic searches of appropriate bibliographic databases were conducted to collate literature involving self-reported experiences of diagnosis of YOD. Eight out of 47 initially-identified papers satisfied our search term criteria, and all papers were quality assessed using Walsh & Browne's criteria for methodological appraisal.

Results: The review of the literature emphasises that delays in diagnosis can often be attributed to (1) delays in accessing help by the younger person, and (2) misattribution of symptoms by the clinician. The impact of diagnosis is influenced by the language used by the clinician, and the younger people diagnosed with dementia and their families' reactions to the diagnosis vary from feelings of reassurance (in that their symptoms are now explained)**,** to shock and destabilisation.

Conclusion: Understanding the perspective of a younger person undergoing assessment for dementia is important both in terms of promoting future engagement with services and in designing and evaluating interventions to improve the diagnostic process. This review suggests that improving the recognition of presenting symptoms, reducing diagnostic errors and identifying the emotional needs arising from diagnosis are required to improve the diagnostic experience for younger adults.

Keywords: young onset dementia, dementia assessment, diagnosis, lived experiences

Background

The process of being assessed and receiving a diagnosis of dementia can be long and daunting (Vernooij-Dassen, 2006). This is particularly true for younger individuals (Svanberg, Spector, & Stott, 2011; D van Vliet et al., 2013) as dementia is typically under-recognised in this age group and often requires more complex investigation. Approximately 42,325 people living in the UK have a diagnosis of YOD (Dementia UK Update, 2014), with an estimated incidence of 11.5 YOD cases per 100,000 people (Mercy, Hodges, Dawson, Barker, & Brayne, 2008).

Young onset dementia (YOD) is typically more complex to diagnose than late onset dementia as presentations are likely to be of rare cause disorders and the common dementias frequently present with atypical symptoms (Rosness, Engedal, & Chemali, 2016; Rossor, Fox, Mummery, Schott, & Warren, 2010; Vieira et al., 2013). The increased frequency of first presentation to services being with symptoms other than memory loss means that misdiagnoses due to other causes, such as psychiatric disorders, depression, and other neurological illness (Vieira et al., 2013), are common. Collectively, this may lead to prolonged delays in receiving a confirmed diagnosis (Draper et al., 2016), significant uncertainty for families and delay in accessing suitable support (Williams, Dearden, & Cameron, 2001) for individuals and their families. These issues could be mitigated by more timely and accurate diagnoses and increased sensitivity from clinicians when discussing and delivering the diagnosis (Millenaar et al., 2016; Sansoni et al., 2016).

Advocates for younger people with dementia **(**YPD) have shared the personal experiences that they encountered during their own diagnosis. These include a prior (mis)diagnosis of

burnout and depression (Rohra, 2016) resulting in unnecessary delay and a 'feeling of unexpectedness' when the diagnosis was formally confirmed (Greenwood & Smith, 2016; Harris, 2004; Roach, Drummond, & Keady, 2016). Rohra describes " (I) *saw myself on a slide that descended into a black tunnel, down and down … I began to cry*" (page 42, Rohra, 2016). On the other hand, Kate Swaffer, who is living with YOD, has argued that an early diagnosis can be regarded as empowering, enabling and assisting the individual to remain independent for longer (Swaffer, 2016).

Despite these highly valuable insights, there is no published evidence which focuses specifically on the personal experiences of younger people who ultimately receive a diagnosis of dementia, with respect to the challenges individuals have faced. Research that has involved younger people living with dementia has predominantly focused on epidemiological studies (Harvey, Skelton-Robinson, & Rossor, 2003) and their clinical or service needs (Mayrhofer, Mathie, McKeown, Bunn, & Goodman, 2018; Roach, Keady, & Bee, 2012). More recently, it has been emphasised that people living with dementia should be given the right to speak about their experiences (Rohra, 2016) and be listened to (Jonas-Simpson, 2003). A greater understanding of the experience younger adults encounter, including identifying challenging barriers during the diagnostic process, could help to provide a more person-centred diagnostic approach for younger adults. This review aims to bring together qualitative self-reports from YPD on the journey to diagnosis to better understand both positive and negative experiences of diagnosis which could shape future guidance about best practice.

This review forms part of the evidence for ongoing research conducted by the authors, aimed at improving the quality of diagnosis for YPD (http://www.ucl.ac.uk/psychiatry/the-angela-project) in a national project called the Angela Project. Importantly, the findings from this review were used to derive a round one questionnaire for a national Delphi consensus study

with people living with YOD and their family supporters on the most effective ways of how to improve the diagnostic process.

The aims of this literature review were to (1) collate the existing research base regarding the self-reported experience of receiving a diagnosis of YOD and (2) to summarise the themes and patterns that emerged.

Method

A comprehensive search of the literature was conducted for a scoping review in May 2017 and then updated in November 2018 using two electronic search engines: PubMed and Web of Science. As this literature search formed part of a Delphi consensus component of a larger study, we had to be very specific about our search terms and selected these two electronic databases due to their specific focus on life sciences and biomedical fields of research. We only included research articles that had been published in peer reviewed journals to ensure the studies used a robust and rigorous evidence base and methodology.

Search Strategy:

Systematic and concise terms were used during the search of relevant papers. A breakdown of the terminology used is summarised in Table 1. The terms in the groups were combined using the AND function in the following manner: 1 AND 2 AND 3 AND 4.

Group	**Terms (in title or abstract)**

1 – defining terms for young onset	TS=(("young onset" OR "younger onset" OR "early onset" OR “presenile” OR "working age" OR “YOD” OR "under 65") NOT (“elderly” OR “older” OR “late”)
2 – defining terms for dementia, and individual diagnoses	TS=("dementia" AND "Alzheimer's" OR "vascular dementia" OR "frontotemporal dementia" OR "Semantic dementia" OR "Huntington’s disease" OR "acquired brain injury" OR "Parkinson’s disease" OR "Creutzfeldt-Jakob" OR "CJD" OR "Lewy bodies" OR "Picks disease" OR "cognitive impairment" OR "neurocognitive disorder" OR “Posterior Cortical Atrophy”)
3 – defining terms for the diagnostic process	TS=(“diagnosis” AND “assessment” OR “diagnostic” OR “GP” OR “misdiagnosis” OR “misdirection” OR “referral”)
4 – defining terms for lived experiences	TS= (“experience” OR “Quality of Life”)

Table 1: **Search terms used for the review.**

Additional inclusions

Papers published until 30th May 2017, with no date restrictions, were included in the comprehensive search. On 15th November 2018, the search was re-run and updated using the

original search terms to establish if any additional papers had been published. Protocol-driven search strategies were supplemented with snowballing methods to search for additional relevant papers (Greenhalgh & Peacock, 2005). These included reference list and citation searches, author searches, and hand searching of key journals.

Exclusion criteria

Papers meeting any of the following criteria were excluded:

- Studies involving non-primary neurodegenerative conditions, such as Korsakoff syndrome or an alcohol induced dementia, and HIV related cognitive impairment, as. individuals with these conditions traditionally have different pathways to care.
- Dementia research studies solely focused on late-onset dementia, as these would not reflect the experiences of people with YOD.
- Articles that were not peer reviewed.

Inclusion criteria

To be included, papers needed to meet all the following criteria:

- Qualitative or mixed method research that directly involved people living with YOD.
- Investigations of the lived experiences of YOD and where themes surrounding diagnosis were reported.
- Research that involved speaking directly to those affected by YOD (i.e. those diagnosed and/or their family members/carers).
- Articles were published in English.

Procedures for Study Selection and Review

Abstracts were retrieved, examined and reviewed for their relevance based on the inclusion criteria by one author (MOM), then considered and discussed during project meetings with two further team members (JC, JP). Full texts of articles were then retrieved and examined by three authors (MOM, JC and JP) in terms of their relevance.

Quality Assessment

A quality checklist with 12 criteria for qualitative research (Walsh & Downe, 2006) was used to appraise the methodological quality of the studies (see Table 2 for the included studies). Papers were scored on 12 criteria. If a criterion was met, the authors rated the papers with 1, while the papers that did not meet a criterion were rated with 0. If a criterion was half-met, the papers were rated with 0.5, and N/A was used if a criterion was not applicable to the given study. This led to a final score out of 12. The quality of the included studies was checked by one rater (MOM). The quality assessment of papers can be found in Table 3.

Results

Search Results

The search identified 47 research articles, of which 11 were included in the in-depth review using the criteria stated above. Eight of the 11 articles met the inclusion criteria. A breakdown of the stages involved in highlighting the appropriate papers for the review is summarised in Figure 1.

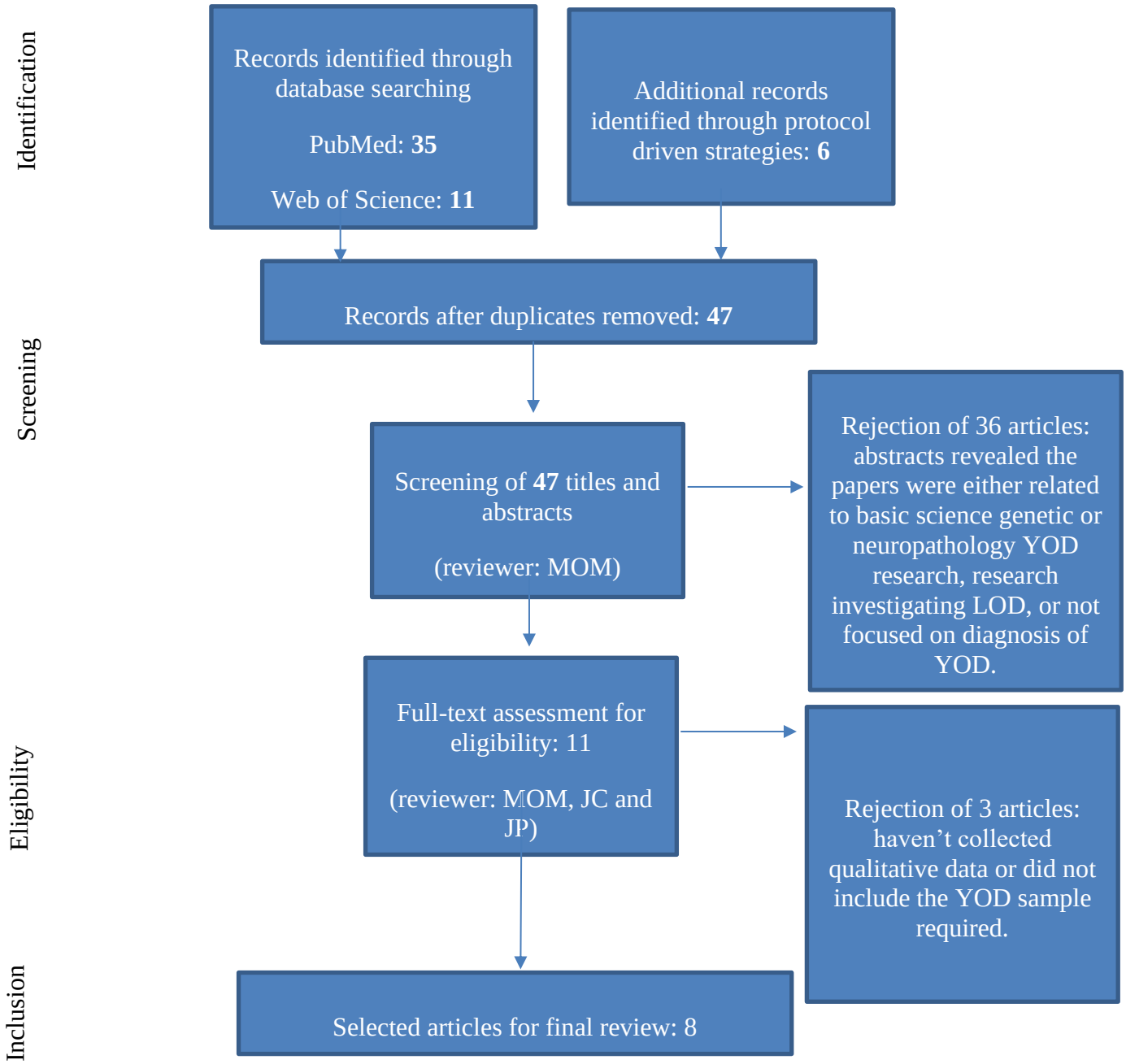

Figure 1: PRISMA flowchart of the literature selection procedure (Moher, Liberati, Tetzlaff, & Altman, 2010).

Table 2 below summarises the studies found in the literature search that met our inclusion and exclusion criteria, providing quotes from those interviewed with a comment on our interpretation of the quotes, and the conclusions made by the authors. The eight studies have been listed alphabetically according to the first author per study.

Author/Year	Country	Study Population	Diagnosis of YPD	Measures	Quotes	Our interpretation of quotes and findings	Conclusions by authors of paper
Beattie, Daker-White, Gilliard, & Means (2004)	UK	PwYOD (14): age: 59.4 years (41-66); gender: 5 females, 9 males	NS	Semi-structured Interviews	"*'I went to [mentions four different hospitals] and had the scans. I went there last November and I went last January, this year, and I should have heard from [the hospitals] in June this year, I should have went, but I haven't heard anything off them at all, right*.' (YP 6) The diagnostic testing was perceived as 'baffling', which left one participant (and carer) feeling incompetent, distressed and	The quote by YP 14 highlights how this particular person felt that the diagnosis was fed back in a "brutal" manner and was perceived as traumatic or unempathic.	The paper emphasises the delays to diagnosis that younger adults experience, the poor quality of information post-diagnosis, and the lack of explanation about next steps.

					unprepared for the receipt of distressing news: *It was very blunt, yeah. It was brutal, that's the only word for it.* (YP 14)'"		
Griffin, Oyebode, & Allen (2016)	UK	PwYOD (5): age: 46 – 62; gender: 2 females, 3 males	BvFTD	Semi-structured Interviews	"*David explained that the people in hospital with him 'had problems with their brains' but then continued to say the doctor had said he 'must have that [frontal-temporal dementia] but I haven't because they do things wrong, but I don't do things wrong'*."	This quote highlighted a difficulty in fully understanding the impact and meaning of the diagnosis by the recipient.	Following a diagnosis of BvFTD, it was noted that there was a lack of emotional reaction and reflection on the diagnosis for one recipient. This may suggest challenges in connecting with the meaning of the information relayed, seeing it as something 'other' and not relevant to own personal

							experience, together with lack of insight. This was particularly when reflecting on the meaning of the dementia diagnosis itself, as well as the new change of lifestyle as a result of the dementia symptoms and diagnosis.
Harris (2004)	USA	PwYOD (23), age 56 (43-68); gender: 13 females, 10 males	14 AD, 6 FTD, 1 MCI, 1 HD.	Focus groups, face-to-face interviews, and on-line interviews	*"People tend not to believe that I have anything wrong because I look and sound so healthy. One of my major stresses was getting my doctor to believe that it's more than stress, depression, or burn out. It's easier now that I don't tell her anything about it I have to get my medicine from my other doctor."* (Participant with AD)	Misdiagnosis is a possible reason why obtaining an accurate diagnosis is challenging.	Difficulty obtaining an accurate diagnosis was a theme found in the reports, reported by 14 of the 23 participants. As captured in the quote, recipients experienced not being believed and lack of knowledge about how dementia presents in younger people by professionals.

Hoppe (2019)	The Netherlands	PwYOD (7) age: NS; gender: 4 females, 3 males. Family members (39), age: NS; gender: NS	NS	Semi-structured interviews	"*I went to the sauna with my sister-in-law and said that I was worried. Yes, we talked about it. That was four years before the diagnosis. In that period I was already worried, and that is now more than six years ago. That was in the beginning and of course you don't want to know.... (Ellen, 55, partner)"*	Recognising the prodromal symptoms and speaking with family members about these can start the journey to diagnosis.	The paper highlights the shift in the meaning of uncertainty in the pre-diagnostic illness trajectory of YOD.
Johannessen & Moeller (2013)	Norway	PwYOD: (20): age: mean 62 years (54-67); gender: 8 females, 12 males	NS	Semi-structured Interviews	"*'It started when I was doing a lot of things wrong at work – I also felt that something was wrong. My colleagues at work experienced it too, as they told me that it seemed as if I was struggling and they asked me if I was tired. I felt tired also. [Number 20]*'	The quote from Number 20 highlights how symptoms can be recognised in the workplace, and by making society more aware of YOD could better support these individuals to	Two main themes emerged from this study: (1) the process towards a dementia diagnosis, with subthemes (i) changes during the prodromal period (ii) and the impact of "being diagnosed; and (2) fighting for dignity

					‘*It was a shock and it scared me, because I know what is to come and it is quite terrible that I will suddenly turn out to be, ugly as it sounds, ‘a second-class citizen’. [Number 4]”*	accessing appropriate help. Number 4, emphasises the issues of present stigma surrounding having a diagnosis of dementia.	after being diagnosed with dementia with subthemes: (i) intrapsychic challenges and (ii) social challenges.
Rabanal, Chatwin, Walker, O’Sullivan & Wilkinson (2018)	UK	14 people with YOD aged between 57 and 67 years	NS	Interviews. Five interviews were paired (person with dementia and their carer), but the carer voices are not reported here.	*“The doctors and consultants were saying, ‘no, you’re too young to have dementia’. At this time I was about 49. So their thinking was it could be depression or stress because I was still working at this point.* (Participant 1)”	Making clinicians more aware of YOD could reduce delays by not misattributing symptoms of dementia as psychiatric disorders.	After seeking medical help, people reported that the GP did not recognise that dementia could be a cause of the symptoms and misattributed the symptoms to a psychiatric disorder.

					"[*They said] it's early onset Alzheimer's disease, just out of the blue like that. Well I was numb. (Participant 3) … it was awful. I felt like I was hit by a sledgehammer… I felt as though somebody had pulled the plug out of everything. (Participant 5)"*	The quote by participant 5 demonstrates the impact of receiving a diagnosis of YOD which can be influenced by how the diagnosis is communicated.	Regardless of how the diagnosis was delivered, there was unanimity over the shock of having the diagnosis confirmed.
Roach, Drummond & Keady (2016)	Canada	Nine families made up of 20 participants, including nine spousal pairs, and two young adult families. PwYOD (9); gender: 0	7 AD 1 PCA 1 Mixed Vascular dementia.	Research Interviews with each of the families on two separate occasions.	*"And again the age factor. And that's the big deal, that nobody would say that this is Alzheimer's or dementia at this age".*	Increasing awareness of YOD can reduce unnecessary delays to receiving a diagnosis and accessing the support needed.	Lack of information during the diagnostic experience and doubt from healthcare professionals about the diagnosis were themes found in their study.

		females, 9 males.					
Wawrzicznyet al (2016)	France	PwYOD (16), gender NS and (16) caregivers, gender 7 females, 8 males	AD	Semi-structured Interviews	"*'Couple 15: I didn't see the onset of the disease because I was always distracted. I remember the catastrophes I caused at my job, especially in times of stress —like when I was going to be evaluated, I would forget a document at home. (person with AD)*' '*Couple 11: I explained what I was feeling, and she [the neurologist] practically told me I'm crazy. (person with AD)*' '*Couple 02: The doctor we had at that time said he's depressed [...]. So, we changed doctors, and my husband said to him, 'Now, I'm panicking, I can't go on, I can't do anything anymore,*	The quotes from couple 15 and couple 11 emphasise a misattribution of symptoms resulting in misdiagnoses. The quote from couple 02 highlights how the lack of recognition of YOD by GPs can result in misdiagnoses resulting in distress for the person.	The 'need to know' the cause of the dementia, and after diagnosis, the 'need not to know more' about the diagnosis for those with YOD were two prominent themes in their study.

					I do any old thing, I forget'. (caregiver)""		

Table 2: Main characteristic of the eight included studies. NS means "not stated". AD = Alzheimer's disease, PCA = Posterior Cortical Atrophy, FTD= Frontotemporal dementia, BvFTD = behavioural variant of frontotemporal dementia, HD = Huntington's disease, MCI = Mild Cognitive Impairment.

Table 3 below shows the Quality Assessment of the included eight studies found in the literature search.

	Beattie (2004)	*Griffin, Oyebode and Allen (2016)*	*Harris (2004)*	*Hoppe (2019)*	*Johannessen & Moeller (2013)*	*Rabanal et al 2018*	*Roach et al 2016*	*Warwrzicny et al (2016)*
Clear statement of, and rationale for, research question, aims and purposes	1	1	1	1	1	1	1	1
Study thoroughly contextualised by existing literature	1	1	1	1	1	1	1	1
Data collection strategy apparent and appropriate	1	1	1	1	0.5	1	1	1
Method/design apparent, and consistent with research interest	1	0.5	1	0.5	0.5	1	1	1
Sample and sampling method appropriate	0	1	0.5	1	0.5	1	1	0
Analytic approach appropriate	1	0.5	0	0.5	1	1	1	1
Context described and taken account of in interpretation	1	1	0.5	1	0.5	1	1	1
Clear audit trial given	1	1	1	1	1	1	0.5	0.5

Data used to support interpretation	1	1	1	1	1	1	1	1
Researcher reflexivity demonstrated	1	1	0.5	0.5	0.5	0.5	0.5	0.5
Demonstration of sensitivity to ethical considerations	1	1	0.5	1	1	0.5	1	1
Relevance and transferability evident	1	1	1	0.5	1	1	1	1
Total Score	11	11	9	10	9.5	11	11	10

Table 3 Quality Assessment of the included studies based on Walsh & Downe's (2006) suggestions. 1 = criterion met, 0.5 = criterion partially met, 0 = criterion not met.

Findings

The research papers found in the literature search were read and cross-validated to explore which similarities, differences and key issues were most frequently reported. Quality Assessment using Walsh and Downe's criteria for appraising qualitative research studies (Table 3; Walsh & Downe, 2006) demonstrated that all papers scored above 9/12 according to their criteria (see Table 3). Through familiarising ourselves with the quotes and interpretations in the highlighted papers, we identified overlapping themes across studies about the experiences of YPD and their supporters and the quality of the diagnostic process. These have been thematically synergised and have been grouped:

The journey to diagnosis – delay in accessing help

Hoppe (2019) illustrated that some individuals and their families initially tried to ignore early prodromal symptoms as they wished to lead life as normal or to not attribute the possible symptoms to dementia and this avoidance may have led to not seeking help for early symptoms: "*But you also ignore it (the dementia), you don't want to see it."* (Manon, 33, Daughter; Hoppe, 2019).

Hoppe used the idiom "ignorance is bliss" to describe why people with YOD may not take action to determine a diagnosis. In other words, although most had an existential awareness of an underlying condition, maintaining uncertainty allowed a sense of hopefulness to remain (Hoppe, 2019).

Griffin et al (2015) emphasised that, for some, inadvertently denying initial symptoms could be a result of a dementia sub-type in which lack of awareness is a characteristic feature. For

example, one participant with behavioural variant fronto-temporal dementia (Bv FTD) did not seem to recognise any changes in his own behaviour and showed a lack of emotional reaction and reflection on his diagnosis of YOD. His feelings, on diagnosis, reflected a sense of bewilderment with an awareness of change not recognised as self, “this is not me”. Here the issue was rather that the individual had difficulty in fully appreciating and understanding the impact of the diagnosis on himself. Failure to recognise altered behaviours, could inadvertently contribute to a delayed diagnosis and access to timely support.

Misattribution of symptoms – delay to diagnosis

Under-recognition of prodromal symptoms as dementia by clinicians was identified in most research papers found in the search. Participants in Harris’ (2004) study suggested that as those with YOD in their study looked physically healthy, this resulted in doctors not taking their concerns as seriously. Complaints of memory problems in healthy older adults are likely to prompt investigation for dementia, but the quotes from Harris’ participants demonstrate that psychiatric disorders were more immediately considered as the primary diagnosis. Furthermore, the reports made by participants in Roach et al (2016) highlight how clinicians and healthcare professionals doubted dementia as a cause, specifically because the person was young.

Two studies (Harris 2004: Rabanal et al., 2018) confirm that misattribution of presenting symptoms as psychiatric disorders, particularly depression or ‘stress’ is common in younger adults.

The participants in Harris (2004) reported their frustration that they had to emphasise to clinicians that their symptoms were more than simply being burnt out:

"One of my major stresses was getting my doctor to believe that it's more than stress, depression, or burn out." (Younger person living with AD; Harris, 2004)

The fact that the individual was still working seemed to influence the clinicians. However, it is not uncommon for dementia in younger people to present with non-cognitive symptoms (e.g. psychosis, mood disturbances, personality changes) and for day-to-day activities and function to be well-preserved.

Conversely, Johannessen and Moeller (2013) highlight how those with YOD had difficulties getting health professionals to listen to what was wrong with them. Symptoms were dismissed despite evidence of difficulties with day-to-day tasks or being noticed by others in the workplace (Johannessen & Möller, 2013).

Communicating diagnosis – impact of diagnosis

Language used

Some of the papers reviewed have highlighted that the communication style some health professionals' use when discussing the diagnostic process, (i.e. tests and investigations) and relaying the diagnosis require greater sensitivity and should be tailored appropriately. For example, the way in which the diagnosis was given to a patient and their supporters lacked empathy and was perceived as traumatic in Beattie et al's (2004) study: "*It was very blunt, yeah. It was brutal, that's the only word for it.*" (YP 14; Beattie et al., 2004). The diagnostic testing was also reported as 'baffling' by the participants in Beattie et al's (2004), so much so that one individual and their carer were left feeling incompetent, distressed, and subsequently unprepared for the receipt of distressing news. In particular, this participant felt the diagnostic assessment lacked in structure, with the assessments starting, "*from being casual to an*

extreme" (YP 14; Beattie et al., 2004), highlighting how the format of assessments and the communication of assessments may need to be more consist, to prevent the people from viewing aspects as unprofessional or baffling.

The reaction to the diagnosis: reassuring, destabilising and shock

Wawrziczny, Pasquier, Ducharme, Kergoat, & Antoine (2016) found that receiving a diagnosis of dementia for a younger person could be reassuring, by ending a period of doubt, but also destabilising, by breaking the equilibrium that had been maintained to date. One person reported that "*as long as they haven't made a diagnosis, me, I say [addressing her spouse with AD], 'Maybe you're depressed, huh?' [...] But then we won't be able to hide anymore, once they've said it."* (caregiver; Wawrziczny et al., 2016). This quote also highlights how knowing the cause of the symptoms can provide a sense of control in managing feelings.

Rabanal et al (2018) concluded that regardless of how the diagnosis was delivered, there was unanimity over the shock of receiving the dementia diagnosis. More so, it was apparent in their study that the amount of supporting documentation and information that participants were given at the point of diagnosis, sometimes caused them to feel overwhelmed, with one participant feeling 'bombarded' with leaflets (Rabanal, Chatwin, Walker, O'Sullivan, & Williamson, 2018). Having the opportunity to see someone face-to-face who can offer emotional support immediately after diagnosis was also highlighted as something that would be highly beneficial.

Discussion

The studies from the literature search, provide important insights into the personal experiences of younger people who receive a diagnosis of dementia. They illustrate how individuals understand and make sense of the changes within themselves, and the impact the diagnosis has on their lives. There is a lack of research focused on the diagnostic experience in younger adults with dementia, with only eight papers found in the literature search where the diagnostic experience was partially discussed.

Under-recognition of and awareness about dementia in younger people, together with misattribution of symptoms are both major contributing factors to the much longer delays in receiving a diagnosis for a younger adult compared to older adults. Statistical reports (Draper et al., 2016), demonstrate a time to diagnosis of 4.4 years in younger adults with dementia, compared with 2.2 years in late onset dementia (LOD) (van Vliet et al., 2013). Williams (2001) highlighted that most younger adults experience chaotic pathways into care, with many seeing 2-5 different specialist consultants before receiving a definitive diagnosis. Although seeing multiple specialist consultants potentially adds to the delay in confirming diagnosis, it could be argued that this is unavoidable when complexity and atypical presentations necessitate extensive investigation and assessment. The reports outlined here, provide insight into delays, suggesting that these can be person, age- and/or diagnosis-specific. Van Vliet et al., (2013) investigated factors related to time to diagnosis for both YOD and LOD. They found that the data relating to the patients' family history of dementia was missing in some individual cases, and that in the majority of cases, the person providing the family history was neither a spouse or a partner so may have been unaware of any family history. Additionally, they found that having a diagnosis of YOD was the most important

predictor of a longer period between symptoms and confirmed diagnosis, and how FTD in particular (regardless of age) was associated with longer durations to diagnosis.

An individual's personal preference to seek help, initial misdiagnosis and lack of recognition of presenting symptoms were common self-reported experiences found in this review.

For example, most individuals presented first to their GP in primary care. It is of concern that many participants in the studies suggested and experienced their GPs and secondary care doctors to be lacking in the knowledge that dementia is a condition that can affect younger people: *"The doctors and consultants were saying, 'no, you're too young to have dementia'. At this time I was about 49 ..."* (Participant 1; Rabanal et al., 2018). Making the diagnosis, or making a decision to refer, is often contingent on enquiry about additional unusual or atypical symptoms, taking a history from an informant, and a knowledge of presenting symptoms that often differ from late onset disease. Lack of familiarity and enquiry of symptoms that may be more typical in rarer dementia types can lead to a dismissal of symptoms. It can also cause a misattribution of the symptoms to primary psychiatric disorders. This can be accompanied, as illustrated here, by frustration for patients and a sense of having to fight to be heard. The high rates of psychiatric misdiagnosis are driven by the significant overlap in symptoms of neurodegenerative disease especially bvFTD and psychiatric disorders (Shinagawa et al., 2014). In one study with large patient sample size, 28% of individuals had a prior incorrect psychiatric diagnosis (Woolley, Khan, Murthy, Miller, & Rankin, 2011). They found that across groups, depressive disorders and bipolar affective disorder were the most frequent misdiagnoses. Also, a diagnosis of schizophrenia was not uncommon. Rates ranged from <12% in those with atypical presentations, e.g. prominent language, speech or movement disorders, and up to 52% in those with bvFTD (Woolley et al., 2011). Over 50% of younger

patients waited up to 3 years before the diagnosis was revised, which therefore results in delays in receiving an accurate diagnosis.

A recently published article by Harding et al (2018) emphasises how younger people with PCA often initially report their first symptoms to their eye health professionals (e.g. optometrists) in the first instance. Ensuring that awareness of rarer YOD is raised beyond GPs in primary care, could led to quicker identification and referral for those with progressive visual impairment that is related to dementia (Harding et al., 2018).

The studies found in the review confirm that communication style, language and clarity of explanation, particularly with regard to the often-numerous tests and procedures required for younger adults, have a significant effect on patient welfare and empowerment.

Thus, a sensitive, collaborative and enabling approach to both assessment and disclosure of the diagnosis is crucial, with ample opportunity for discussion as and when an individual feels ready to know more. The complexity of judging how much a person and their family wish to know about the diagnosis has been echoed in other qualitative studies of all-age dementia. Information may be empowering for some, but rejected by others who may choose not to know' (Bunn et al., 2012). The National Institute of Clinical Excellence (NICE) states that "people should be told their diagnosis as clearly and honestly as possible", to "plan effectively for their future" and access treatment and support (National Collaborating Centre for Mental Health, 2007). The personal experience of YPD quoted here emphasises that there are gaps in addressing people's cognitive needs for information and emotional needs for empathy. It is inevitable that addressing a younger person's needs for information about their complex assessment, understanding their emotional reactions to the diagnosis, providing them with information about the future, and building a meaningful life with dementia would

differ depending on the age of onset and the specific dementia sub-type. No practical guidance regarding communication of diagnosis in younger people currently exists.

Importantly, an apparent lack of overt initial reaction may not necessarily reflect the severity of impact that the diagnosis has on the individual. Ensuring that immediate emotional support is available was regarded as more beneficial in the short-term than providing information leaflets on dementia and possibly post-diagnostic support services. A template for good practice in diagnosis has been developed by the Young Dementia Network (https://www.youngdementiauk.org/young-onset-dementia-pathwaywhich) and involves a fresh approach to provide continuity of care and support at and before diagnosis through the provision of a specialist key worker. The YDN recommend that the key worker should (a) have specialist knowledge, skills and experience of the impact on YOD, including knowledge and understanding of rarer forms of dementia, as well as the impact of diagnosis at a younger age for the person diagnosed and their family. They should also (b) provide information, practical and emotional support for family members and (c) provide continuity of support, and enable the person living with YOD to understand their condition and actively engage in their care plan and journey. The key worker should also (d) act as a coordinator of services, organisations and people, helping to connect the person living with YOD to local group and they should be available through a variety of organisations depending on local infrastructure such as primary care, voluntary sector or be based within local mental health teams or neurology services. Providing this initial support through a key worker would provide the needed emotional support immediately following diagnosis that was highlighted as an issue in this literature search.

The British Psychological Society (Watts, McCabe, & Guss, 2018), has recently released advice for clinicians on how best to communicate a diagnosis of dementia, recommending that it be person-centred, taking account of an individual's expectations, preparedness and expressed wishes, and consent should be sought about these. Although these suggestions apply broadly to dementia across all ages, ensuring these principles are considered when diagnosing younger adults may be more challenging. For example, language variants, visuospatial variants and behavioural change are more common in younger people and understanding of the user perspective can be hard to gain. Dementia Australia have published helpful guidance on better ways to communicate more generally with younger people with dementia (Dementia Australia, 2017), including developing strategies to improve understanding and seeking ways to encourage communication and expression when language and communication are often more significantly affected. Whilst this guidance by Dementia Australia does not relate specifically to the diagnostic journey, it does emphasise key considerations that are specific for younger people with dementia.

Additionally, good practice in pre-assessment counselling (La Fontaine, Buckell, Knibbs, & Palfrey, 2014) and establishing informed consent sets the scene for successful future engagement with services – it improves the dialogue between those undergoing assessment and the health professional conducting the assessment, and could support the impact of diagnosis for if and when it is communicated. Informed consent also enables people to adapt in such a way that the advantages of early diagnosis are maximised. People with dementia have explicitly stated that they have the right of a timely and accurate diagnosis (Dementia Action Alliance, 2018), and this is essential for people who are still of working age, with family commitments and financial concerns.

Drawing on the self-reported experiences of receiving a diagnosis of YOD was the purpose of this literature review. Only studies that directly involved the person living with the diagnosis of YOD were included in this literature review. This said, we did include studies that encapsulated the family's experience, where the family member living with YOD was also included (e.g. Hoppe, 2019). The idiom "ignorance is bliss" was used by Hoppe to describe the family supporters' reports that they wanted to ignore the dementia and not see it. Roach, Keady, Bee & Williams (2014), proposed four family storylines to conceptualise interviewed families experiences of YOD - an agreeing storyline, a colluding storyline, a conflicting storyline, and a fabricating storyline (Pamela Roach, Keady, Bee, & Williams, 2014). These storylines and approaches in how families cope post-diagnosis, also illuminates the differing coping strategies pre-diagnosis when faced with behaviours that are not attributed to a known cause. Van Vliet et al., (2011) interviewed family supporters of people living with YOD and reported the long and difficult period they experienced pre-diagnosis. Van Vliet et al., (2011) also found that non-responsiveness from the GP and misdiagnosis were key themes that family supporters reported (Van Vliet et al., 2011). Including the voices of family supporters, as well as the person living with the diagnosis of YOD, offers a unique opportunity to explore similarity and differences between the two different experiences of the diagnostic journey.

Finally, it is perhaps worrying that although two of the articles found in the literature search were published more than 10 years ago (Beattie, Daker-White, Gilliard, & Means, 2004; Harris, 2004) the themes identified remain consistent with more recent reports (Griffin et al, 2015; Wawrziczny et al., 2016), emphasising the recurrence and persisting nature of the issues raised. Furthermore, the research described was conducted in a variety of European countries and North America indicating that the issues highlighted are universal and could benefit from international consensus.

Implications for Research and Practice

Practice

These valuable insights into the personal experiences of receiving a diagnosis of YOD have significant implications for health care professionals working in the field. Implicit within these findings is the need for increased awareness of the impact a diagnosis of dementia has on younger adults in relation to their work, relationships and personal circumstances. It is hoped that reducing delays and avoiding misdiagnoses could potentially lead to a better adjustment to the diagnosis of dementia.

Improving recognition of symptoms through the education of key professionals, clarifying pathways into care and identifying local experts with specialist expertise remain key goals. In 2018, the Young Dementia Network in the UK, produced a diagnostic algorithm to support General Practioners (GPs), (https://www.youngdementiauk.org/gp-decision-making-guide) in identifying possible early symptoms of YOD, and the steps and checks GPs should undertake. Similarly, a YOD care pathway (https://www.youngdementiauk.org/young-onset-dementia-pathway) bringing together the experience and expertise of people affected by YOD and the people who work with and support them, identified a series of recommendations to guide policy makers, commissioners and practitioners in designing and delivering services which empower and meet the needs of younger people with dementia and their families. The pathway identifies key standards about the process from pre-diagnosis and transition to end of life care.

Research

Although there is a diagnostic algorithm to support GPs in making identifying possible YOD, currently no such guidelines about best practice in diagnosis of YOD exist for secondary care clinicians**.** General guidelines for dementia (e.g. NICE) are presently not informed by the lived experience from those directly affected. While most clinicians would agree that a truly collaborative and person-centred approach to diagnosis is crucial, limited evidence is available to support how this could be best achieved. YPD have specific needs, arising from their stage of life and the consequence of diagnosis at a younger age has a devastating impact on life trajectory, relationships and role.

Putting the YPD at the centre of this process to understand what works best to improve the patient experience would mark an advance in approach and policy. A deeper enquiry into the experience of receiving a diagnosis of dementia in younger adults, where both people with dementia and their families are included is necessary (Stamou et al., 2018). A consensus Delphi study of people living with YOD, and their families is the current focus of our own research, where our goal is to capture experiences of the diagnostic process from YPD and their supporters, and most importantly use their expertise to inform how and where improvements should be made. This review has provided theoretical and methodological guidance in steering the focus of questions in the first round of that Delphi study.

Strengths and limitations

This review is the first to focus specifically on qualitative accounts of the diagnostic journey for younger people living with dementia. Importantly, this review forms the basis of a UK national study asking people with YOD and family supporters about their experiences during the diagnostic process and provides insight into the key areas of concern that have been

reported in earlier studies. It is important to note that this literature review was conducted to inform the first round of open-ended questions for a unique and innovative national Delphi study which asked for the opinions of those living with YOD and their family supporters, alongside the perspectives of experts in the field. We were particularly interested in exploring if there were any qualitative studies that purely focused on the diagnostic journey, and found there were none. In addition, we were interested in the key topics of interest relating to the diagnostic journey for younger people with dementia that emerged from the search. Due to the scope of the current study, we were limited in the appropriateness of the databases we accessed as part of this thematic review; however, further reviews in the future could consider re- running the search across a broader range of databases.

Conclusion

In conclusion, the literature highlights that there is scope to improve the process surrounding diagnosis for YPD, particularly in relation to the journey to diagnosis, misattribution of symptoms and communicating the diagnosis. The Angela Project, funded by the Alzheimer's Society, is a current project which focuses on improving the quality and accuracy of diagnosis and support and services for YOD. This review has illustrated the importance of the voice of YPD and their supporters in shaping good practice.

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