

## EDITORIAL

# Young-onset dementia: not the same as late-onset dementia – highlighting the differences in diagnosis, care, treatment, and models of care

The rising prevalence of dementia includes an increase in people with young-onset dementia (YOD) (Hendriks *et al.*, 2021). YOD is a growing public health concern often overshadowed by its more prevalent and well-known late-onset dementia (LOD) counterpart. YOD refers to a dementia where the first symptom onset occurs when the person is less than 65 years of age. Though less common than LOD, YOD is often more challenging to diagnose and manage due to its heterogeneous presentation (Koedam *et al.*, 2009). Misdiagnosis and delayed diagnosis, with up to five years delay, remain major hurdles (Draper *et al.*, 2016; Loi *et al.*, 2022; van Vliet *et al.*, 2013). Additionally, the psychosocial, medical, and policy implications of YOD are distinct from LODs in many ways. Existing support systems for people with dementia are designed for, and predominantly delivered to, older populations. These often fail to meet the specific needs of younger individuals and their families, whose unique challenges necessitate focused effort. In the Oceanic context, a recent paper led by editors Goh, Cations, and Velakoulis highlighted the importance of addressing the specific issues in the YOD population (Cations *et al.*, 2021) and outlined pressing priorities for research, policy, service, and advocacy with significant potential to improve outcomes for people with YOD and their care partners and families.

This special issue in YOD addresses these differences, now with a global perspective. Much has happened in the field of YOD since the last *International Psychogeriatrics* special issue on YOD was published in 2014, guest edited by Koopmans and Rosness (2014). The field has advanced rapidly in the past nine years, including exciting developments in the understanding of YOD pathogenesis, biomarkers and imaging, genetics, cognitive science, clinical trials, and post-diagnosis care. The articles in this collection provide a critical update on YOD research and care. As guest editors for this issue, we hope this special issue will have an international impact on clinical practice, training, research, and policy and raise awareness and

advocate for progress in the YOD field. To this end, we have focused on the aim of improving the lives of people impacted by YOD, via highlighting cutting-edge, high-quality research with diverse methodologies and disciplines that advances and improves knowledge, develops interventions, improves diagnosis and care, and/or informs practice or policy. Based on the submissions for this special issue, we summarize here key targets for focused research attention to promote the well-being of people with YOD and their families.

### Call to action 1: YOD research must be more globally linked to truly represent and meet the needs of the population of people with YOD

Information on the prevalence and incidence of YOD is imprecise and scarce. A recent systematic review and meta-analysis included a total of 95 unique studies investigating the prevalence of YOD (Hendriks *et al.*, 2021). Age-standardized prevalence estimates increased from 1.1 per 100,000 population in the group aged 30–34 years to 77.4 per 100,000 population in the group aged 60–64 years. This gives an overall global age-standardized prevalence of 119.0 per 100,000 population in the age range of 30–64 years, corresponding to 3.9 million people aged 30–64 years living with YOD in the world. A similar review of studies of the incidence of YOD included 61 articles (Hendriks *et al.*, 2023). Global age-standardized incidence rates increased from 0.17 per 100,000 in age 30–34 years to 5.14 per 100,000 in age 60–64 years, giving a global total age-standardized incidence rate of 11 per 100,000 in age 30–64. This corresponds to 370,000 new YOD cases annually worldwide. The incidence of YOD found the majority of studies were from Europe (34 studies), North America (10 studies), Asia (10 studies), Oceania (2 studies), and South America (5 studies). No incidence studies on YOD were found for Africa. Due to insufficient data from studies outside Europe, no subgroup analysis on ethnicity could be performed.

These pooled data mask intra-regional variability that is known to exist in YOD prevalence and incidence. For example, prevalence rates for YOD in minority ethnic communities are higher than for the population as a whole. In Australia, First Nations populations (representing less than 3% of the population) develop YOD at three times the rate of non-Indigenous populations (Radford *et al.*, 2015). More data are needed for global rates in YOD, with attention given to diverse cohorts (including diversity in cultural and linguistic backgrounds and minority groups). For example, published data indicate that LOD rates are 22% higher among black people in the United Kingdom compared with white people, while black and South Asian dementia patients die younger and sooner after diagnosis (Mukadam *et al.*, 2023).

Considering diversity is important because of disparities in the inclusion of minority ethnic populations in dementia research and the known barriers to diagnosis and effective post-diagnosis care. Minority groups face numerous barriers to accessing dementia services, including barriers related to stigma, and healthcare system-related barriers (Cooper *et al.*, 2010; Mukadam, *et al.*, 2011). Potential language and educational differences are also likely to reduce the accuracy of medical, cognitive, and behavioral assessments.

Articles and commentary within this special issue are from the United Kingdom, New Zealand, Netherlands, Australia, and the USA. While this represents intercontinental researchers (three continents of the seven), all these countries are classified as high-income (for the current 2024 fiscal year calculated using the World Bank Atlas method), and all countries either have English as the official language or have an extremely high proportion of English speakers (the Netherlands). Despite the call for articles on this issue being widely shared and specific targeting of research groups from more diverse countries, the articles within are unfortunately not from low- and middle-income countries or from non-English speaking countries. This gap is emblematic of a wider disparity in the availability of research from these regions.

### Call to action 1

*We advocate for active efforts to engage and link research and clinical care teams from all countries and regions. Diversifying YOD research will have numerous benefits, including the development of culturally appropriate, effective, and equitable assessment methods and interventions, ensuring that all populations benefit from research efforts, identifying interventions that are recommended or contraindicated in certain populations, building research capacity in all regions of the world, and*

*ensuring research practices and services that benefit both the scientific community and the population that is actually affected by YOD.*

### Call to action 2: YOD research must be adequately funded and involve all people involved in providing and receiving care, to promote innovation and rapid advancements

Within this issue of original research articles, topics range from emotion recognition, psychotic symptom identification, behavioral symptoms, the dynamics and experiences of spouses and carers, service provision, and grief and loss. These topics touch on experiences spanning pre-diagnosis to end of life and with diverse methodologies including retrospective and prospective qualitative, quantitative, and mixed methods approaches. As guest editors for this special issue, we were struck with the innovation and quality of the work submitted for inclusion.

Sufficient funding to continue and advance this high-quality and diverse research is key to advancing YOD science. Examples of the major benefits of a strong YOD research infrastructure exist. One such case is the Netherlands, where a well-funded YOD research program is integrated into clinical practice to conduct innovative and impactful biomedical and psychosocial (Bakker *et al.*, 2022). For many years, the Netherlands has funded YOD special care units in long-term care facilities that collaborate in the Dutch YOD Knowledge Center. One of the achievements of this knowledge center is that it issued a YOD research agenda with a broad area of topics to be addressed in the upcoming years. This agenda helped the Dutch government decide to prioritize YOD in their national dementia strategy that has a funding budget of 140 million euro (2023–2031). Additionally, in 2022, a national consortium consisting of 12 partners including the Dutch Alzheimer Society was awarded a grant of 6 million euro for research on YOD. This funding will establish a national longitudinal cohort with at least 1000 participants, using innovative approaches to ensure inclusion (e.g., persons with YOD and their carers can include themselves via a digital portal, as a form of “patient-initiated” inclusion). There will be workstreams focusing on advancing research on genetics, postmortem MRI, improvement of the clinical recognition of YOD syndromes and early symptoms, creating a universal clinical tool kit for memory clinics, and studying fitness to drive with a focus on social cognition. Projects will improve diagnostic and post-diagnostic care and support and improve the accessibility and connection between

different types of care and support throughout the caregiving trajectory.

A different example of an effective, integrated YOD research program is available in Australia at the Neuropsychiatry Centre, Royal Melbourne Hospital. Clinical research has been embedded into the clinical program such that the majority of YOD patients and carers will participate in research and consent to the use of clinically derived data for research purposes. This program of clinical research spans the breadth of research domains, from biomarker to imaging, cognitive, genetic, and psychosocial work (e.g., Loi *et al.*, 2022).

Traditionally, medical research has been driven by academics and researchers. However, the landscape is shifting toward a more collaborative approach. In YOD research, incorporating industry, policymakers, and the YOD community as research partners holds immense potential for advancing the field. A collaborative approach harnesses expertise, experience, and insights from diverse communities, ensuring cutting-edge research that focuses on the most pressing issues and priorities that are ethical, relevant, culturally appropriate, and address the specific needs of the YOD population and their family members. Collaboration also aids in recruitment and retention in research and in disseminating research findings in a way that resonates with the community.

### Call to action 2

*Increased funding for collaborative research dedicated to YOD is crucial, which will have long-term benefits and a significant return on investment. There is a breadth of expertise available, from early career researchers to more established researchers, clinician researchers, and policy experts, and this pipeline should be nurtured. For example, recent advances in diagnostics would not have occurred so rapidly within the last decade without significant investment in biotechnology and participation of the YOD community. Strategies should be strategically employed to increase funding, including raising awareness and public support, engaging with and lobbying policymakers and grant makers (diversifying funding from government, corporate, and philanthropic funding), integrating health services and research clinics, and nurturing genuine partnerships and collaboration with diverse groups, including industry and the people impacted by YOD.*

### Call to action 3: there needs to be agreement in the research community upon the nomenclature of YOD

At the time of the previous editorial in 2014, there was no clear consensus on the nomenclature and

operational definition of YOD. A workshop on behalf of the International Psychogeriatric Association (IPA) suggested to use the term “young-onset dementia.” In 2021, an integrative review included 89 papers on YOD and found the following terms used: presenile dementia, early onset dementia, adult-onset dementia, younger onset dementia, and young-onset dementia (van de Veen *et al.*, 2021). “Young-onset dementia” was the most used term in the last two decades; the age of 65 years at first symptom onset was used most frequently to define a cutoff age, and a total of 251 different etiologies were identified. As a follow-up, a Delphi study included 44 experts in the field that reached full consensus on using YOD as the preferred term, and provisional consensus was reached on using the age of 65 at first symptom onset (van de Veen *et al.*, 2022). However, submissions for this special issue indicate that there continue to be various terms used to describe YOD in the literature and in practice, for example, with many authors preferring the term “early onset” dementia.

### Call to action 3

*We call for efforts to disseminate, promote, and implement consistent nomenclature and continued ongoing discussions and refinement of terminology, operational definitions, and implications for use in research and clinical practice as the field advances. This will reduce heterogeneity, ambiguity, and confusion about the criteria and the inclusion of various etiologies in diagnosis and inclusion in clinical trials and help to enhance and compare research. Clear definitions are beneficial for discussions, and decision-making becomes more efficient for conducting research and for clinicians, for example, to develop policies concerning the organization of age-appropriate post-diagnostic services. Shared terminology also optimizes collaboration. By ensuring clear communication and collaboration, consensus terminology will lead to better overall outcomes in the YOD field.*

### Call to action 4: post-diagnostic care in YOD needs evidence to inform policy and services

People with YOD live on average 10 years after their diagnosis, though life expectancy varies considerably depending on dementia type, presentation, and other factors (Gerritsen *et al.*, 2019). The heterogeneity in YOD presentation, etiology, and prognosis makes the funding and provision of post-diagnosis care highly complex. Adding to this complexity is the low prevalence of YOD (relative to later onset dementias), such that people with YOD may be disparately located and professionals may interact



with people with YOD only infrequently. The result is a high level of unmet need among people with YOD and their families during their post-diagnosis illness (Carter, *et al.*, 2018; Cations *et al.*, 2017). High-quality research focused on post-diagnosis YOD care is vital to meet this unmet need. Research in YOD (so far) tends to disproportionately favor biomarker, imaging, and other studies aimed at understanding and diagnosing YOD. While the pre-diagnosis and diagnosis phases remain important research targets, post-diagnostic issues should also be based on the best evidence for effectiveness and feasibility for real-world delivery. Research that guides policy is lacking, and funders require data about the cost-effectiveness and adaptability of services and models of care within local markets (Carter *et al.*, 2018; Cations *et al.*, 2021). At present, the delivery of specialized services is heavily reliant on the availability of individual champions within individual services (Bakker *et al.*, 2022). Access to these services is therefore inequitable.

#### Call to action 4

*A stronger evidence base must be built to guide post-diagnosis management, care, and support for people with YOD. Current priorities for research investment include trials of non-pharmacological and rehabilitative therapies for YOD (e.g., cognitive stimulation, dyadic occupational therapy interventions, exercise interventions), research to inform driving safety and cessation interventions, psychological therapies to promote mental well-being and reduce suicide risk, and strategies to prolong engagement in paid and unpaid work. Research focused at the system level will also be vitally important for guiding policy making, with a focus on implementation outcomes including the acceptability, cost-effectiveness, and feasibility of delivering specialized services across regions.*

#### Call to action 5: more needs to be known about the palliative care needs of people with YOD

Palliative care in people with YOD remains a relatively undiscovered area with several differences between YOD and LOD, including the impact of comorbidity, survival, pharmacotherapy, and advance care planning (Koopmans *et al.*, 2015). Despite what is known about the prognosis and life expectancy in YOD (e.g., Gerritsen *et al.*, 2019), much remains to be discovered about how best to support people with YOD and their families regarding palliative care. Two recent qualitative research papers investigated preferences regarding advance care planning in people with YOD and their family caregivers (van Rickstal *et al.*, 2019, 2022). Family caregivers feel limited engagement in advance care planning, do not consider it useful, emotionally

protect themselves, and prefer a day-to-day attitude. Persons with YOD lacked awareness of the concept of advance care planning and had barely engaged in future planning. A recent Dutch study on people with YOD living in residential care confirmed that there is still a lot of work to be done to start conversations about advance care planning (Maters *et al.*, 2024). Only 5.4% of the residents had a written advance directive, and a quarter of the residents had written documentation of the former general practitioner.

#### Call to action 5

*There is a dearth of studies on palliative care and end-of-life preferences in people with YOD, even though their needs (and their family members' and carers' needs) are likely to be distinctly different from those with later onset dementia. This gap in knowledge hinders us from providing optimal care for the YOD community, who also have the right to a dignified death according to their wishes and preferences. We call for more funding and more studies on the differences in palliative care approaches, advance care planning, and symptom management between YOD and LOD, to ultimately lead to better palliative care practices for YOD, including improved symptom management, enhanced communication, and overall better quality of life at the end of life.*

#### Summary

Our call-to-action items are highlighted by the articles within this special issue. Indeed, the articles have met the aim of the issue, highlighting that YOD is not the same as later onset dementia, with key and meaningful differences in diagnosis, treatment, and models of care. We have proposed solutions and encouraged specific actions from researchers to raise awareness and maintain progress in the field. We believe that addressing the highlighted call-to-action items will result in a thriving research ecosystem for YOD, leading to diagnostic advancements, reducing stigma, more accessible and equitable treatment options, and improved psychosocial and care support from pre-diagnosis to end-of-life care. We hope you enjoy this special issue, with a renewed enthusiasm and understanding of the critical need to ensure YOD receives the focused attention it deserves. Ultimately, a thriving YOD field will significantly enhance the quality of life for individuals living with YOD and their families, optimizing outcomes, and delivering economic and social benefits.

#### Conflict of interest

None.

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