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Carer burden and behavioral disturbance is similar between younger-onset Alzheimer's disease and behavioral variant frontotemporal dementia

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ABSTRACT

Objectives: Carer burden is common in younger-onset dementia (YOD), often due to the difficulty of navigating services often designed for older people with dementia. Compared to Alzheimer's disease (AD), the burden is reported to be higher in behavioral variant frontotemporal dementia (bvFTD). However, there is little literature comparing carer burden specifically in YOD. This study hypothesized that carer burden in bvFTD would be higher than in AD.

Design: Retrospective cross-sectional study.

Setting: Tertiary neuropsychiatry service in Victoria, Australia.

Participants: Patient-carer dyads with YOD.

Measurements: We collected patient data, including behaviors using the Cambridge Behavioral Inventory-Revised (CBI-R). Carer burden was rated using the Zarit Burden Inventory-short version (ZBI-12). Descriptive statistics and Mann-Whitney U tests were used to analyze the data.

Results: Carers reported high burden (ZBI-12 mean score = 17.2, SD = 10.5), with no significant difference in burden between younger-onset AD and bvFTD. CBI-R stereotypic and motor behaviors, CBI-R everyday skills, and total NUCOG scores differed between the two groups. There was no significant difference in the rest of the CBI-R subcategories, including the behavior-related domains.

Conclusion: Carers of YOD face high burden and are managing significant challenging behaviors. We found no difference in carer burden between younger-onset AD and bvFTD. This could be due to similarities in the two subtypes in terms of abnormal behavior, motivation, and self-care as measured on CBI-R, contrary to previous literature. Clinicians should screen for carer burden and associated factors including behavioral symptoms in YOD syndromes, as they may contribute to carer burden regardless of the type.

Key words: carers, young onset dementia, Alzheimer's disease, frontotemporal dementia

Background

Younger-onset dementia (YOD), defined as dementia with symptom onset before 65 years of age (Rossor *et al.*, 2010), accounts for 7–9% of all dementias (Hendriks *et al.*, 2021; Ward *et al.*, 2022).

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The majority of carers of people with YOD face significant burden (Dixit *et al.*, 2021; Lim *et al.*, 2018) and adverse mental health (Kang *et al.*, 2022) due to diagnostic delay (Loi *et al.*, 2020), role changes, social isolation and lack of appropriate support (Cations *et al.*, 2017; Sansoni *et al.*, 2016). Increased burden for carers of people with YOD has been attributed to the exacerbation of financial and life stressors on the carer (Hall and Sikes, 2017; Roach and Drummond, 2014), the increased length of time in the caring role (Chiari

et al., 2021; Kang *et al.*, 2022), and strain on the family system (Cations *et al.*, 2021). There have recently been calls from Australia and Europe for the prioritization of investments in YOD carer research and support (Cations *et al.*, 2021; Metcalfe *et al.*, 2019).

While carer burden may be higher in people supporting those with YOD compared to older-onset dementia (Kimura *et al.*, 2021; Lim *et al.*, 2018), there is scant literature available about the impact of the YOD subtype on carer burden. Comparing Alzheimer's disease (AD) with behavioral variant frontotemporal dementia (bvFTD), including both younger-onset and older-onset, carers of those with bvFTD are reported to have higher burden (Liu *et al.*, 2018; Mioshi *et al.*, 2013; Wong *et al.*, 2012). This has been attributed to the behavior-related symptoms in bvFTD, such as apathy, depression, impulsivity and disinhibition (Mioshi *et al.*, 2013; Wong and Wallhagen, 2014; de Vugt *et al.*, 2006).

Yet younger-onset AD is associated with greater clinical heterogeneity due to greater dysexecutive and behavioral disturbances (Koedam *et al.*, 2010; Sirkis *et al.*, 2022), which might also drive increased carer burden. One study found a trend that carers of people who have bvFTD may experience higher levels of carer burden compared to carers of those with younger-onset AD (Uflacker *et al.*, 2016). This may be exacerbated by the lack of dedicated support or resources available for people with YOD and their families to help them understand, cope and deal with the disease and its various impacts on their lives (Cations *et al.*, 2017, 2021; Sansoni *et al.*, 2016).

The consequences of high carer burden may subsequently accelerate placement of younger people with dementia into an aged care system that is ill-equipped for their specific needs (Yaffe *et al.*, 2002). A better understanding of carer burden in different subtypes of YOD will help services and policy-makers identify and support at-risk carers and develop interventions to decrease burden.

This study aimed to explore carer burden in younger-onset AD and bvFTD, testing the view that carers of bvFTD may have higher burden than carers who support people with younger-onset AD. We also hypothesized that carers of younger-onset bvFTD would report more behavioral disturbance than carers of younger-onset AD.

Methods

Study setting

This was a retrospective cross-sectional study of patient-carer dyads assessed at Neuropsychiatry, Royal Melbourne Hospital, Victoria, between 2016 and 2020. Neuropsychiatry is a tertiary assessment service for individuals with neuropsychiatric disorders including YOD, offering both inpatient and outpatient assessments. The service admitted approximately 80 patients per year during the study period, with 31 (38%) people being diagnosed with dementia. Patients with YOD are offered follow-up care in the service's state-wide YOD Clinic, that sees approximately 400–500 patients per year (Loi *et al.*, 2022).

All patients received multidisciplinary including medical and allied health (neuropsychology, occupational therapy and speech therapy) and multimodal assessments. Diagnoses were based on the National Institute of Aging-Alzheimer's Association Criteria (Hyman *et al.*, 2012) for AD and the Rascovsky criteria (2011) for bvFTD.

Participants, recruitment & ethics

All primary, informal carers who looked after patients with younger-onset AD or younger-onset bvFTD subtype were identified as potential participants. YOD was defined as symptoms onset before the age of 65 years. In addition, carers were defined as a spouse, relative or friend who identified as the primary carer of the person with YOD. Both inpatients and outpatients were eligible to participate in the study. As the questionnaires were in English, this limited participants who were not fluent in English.

Carers who consented to study procedures, completed questionnaires as part of a larger ethically approved study, *Biomarkers in younger-onset neurocognitive disorders* (BeYOND study) during their clinical Neuropsychiatric admission (Loi *et al.*, 2021) (Melbourne Health Research Ethics Committee 2016.038 and 2018.371).

Measures

We collected demographic information about the carers, such as their age at the patient's assessment, sex, relationship with the patient and whether they lived with the patient with dementia. We also obtained demographic and clinical information about the patient, including age at the assessment, sex, past psychiatric history, family history of dementia and duration of dementia symptoms. We used validated questionnaires to obtain carer burden, patient cognition and behaviors listed below.

Carer burden: We measured carer burden using the Zarit Burden Inventory-short version (ZBI). The ZBI-short version is a 12-item questionnaire measuring subjective burden with a 4-point Likert scale. It has a total score of 48 points, with higher scores reflecting higher levels of burden. A score of 17 or more indicates high burden (Bédard *et al.*, 2001).

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Behavioral change in patients: Behavioral changes were evaluated with the Cambridge Behavioral Inventory-Revised (CBI-R) (Wear *et al.*, 2008). The CBI-R is a caregiver-based questionnaire comprising 45 questions divided into ten domains: memory and orientation; everyday skills; self-care; abnormal behavior; mood; beliefs; eating habits; sleep; stereotypic and motor behaviors; and motivation. It is completed by a family member or close friend of the patient, using a Likert scale to describe the frequency of behavior over the previous month where 0 = never and 4 = constantly. The total score is 180, where higher scores indicate severe behavioral disturbance.

Patient cognition: The Neuropsychiatry Unit Cognitive Assessment tool (NUCOG) is a reliable and valid measure of the five cognitive domains (attention, visuospatial, memory, executive function and language). It has a total score of 100, with a higher score indicating better cognition (Walterfang *et al.*, 2006). Scores above 80 are considered normal. However, some patients were unable to complete the NUCOG due to their severe cognitive impairment. In these cases, clinicians completed Mini-Mental State Examination (MMSE) (Folstein *et al.*, 1975). A NUCOG "equivalent" total score was calculated from the MMSE total score for these patients using a formula obtained from the original NUCOG authors (Walterfang and Velakoulis personal communication). This was based on data that compared 562 subjects with MMSE and NUCOG, with the correlation r^2 being 0.91. The information about these participants' MMSE scores and calculated NUCOG scores are in Table 4.

Data analysis

Statistical analysis was performed using Jamovi v1.6 (Jamovi Project, 2021) and SPSS v24 (IBM Corp, 2016). For all demographic variables, variance homogeneity and normality of distribution was tested with the Shapiro-Wilk test. Continuous variables were reported as mean values ± standard deviation (SD). In addition, for the demographic variables, continuous and categorical variables were tested for association with the Mann-Whitney U test and Fisher's exact test, respectively. We used Mann-Whitney U tests and general linear models (GLMs) to compare the differences in CB and other clinical variables between bvFTD and AD. Given the small sample size, inferences for clinical and carer burden were performed using bias-corrected and accelerated (BCa) confidence intervals, computed for all GLMs via nonparametric bootstrapping (1000 replicates). Statistical significance was defined as any confidence interval not capturing the null hypothesis value (at the 95% level).

We estimated GLMs as a post hoc analyses to compare differences in carer burden between the dementia subtypes with total NUCOG score and total CBI-R score, indirect measures of dementia severity, as separate covariates. Furthermore, we also examined the association between behavioral disturbance (CBI-R) and carer burden (ZBI) using Spearman correlation, using Bonferroni correction due to the presence of multiple comparisons.

Results

Demographics of carers and patients (Tables 1 and 2)

Of the 67 carers included in the study, 38 did not complete or partially completed the questionnaires. Four were excluded as they were diagnosed with older-onset dementia. The remaining 33 carers' mean age was 53.8 years (SD = 12.5), with the majority being female (21 females, 64%). For the carers of patients with AD, their mean age was 53.6 (SD = 11.7), with the majority being female (55%). For the carers of patients with bvFTD, their age was similar to AD (mean = 54.2, SD = 14.1), with the majority also female (77%). There was no statistical difference in their demographics.

For the 20 patients with AD, their mean age was 58.1 (SD = 4.78), with 30% of them being female. For the 13 patients with bvFTD, their mean age was 55.8 (SD = 10.4), with 38% being female.

Carer burden (Figure 1 and Table 3)

Using the ZBI-12, the overall mean burden score was 17.2 (SD = 10.5). More than half of the carers of people with AD (55%, 11/20) experienced high burden as defined as a score of 17 or more (mean = 18.5), while over a third of carers of people with bvFTD (38%, 5/13) reported high burden (mean = 15.2). However, carer burden did not differ significantly between AD and bvFTD (mean difference = 2.6, 95%CI [− 5.0, 10.4]).

Clinical variables of YOD (Table 3)

Compared to patients with AD, patients with bvFTD were more likely to have difficulty with stereotypic and motor behaviors (mean difference = 3.9, 95%CI [0.8, 7.0]). In contrast, patients with AD were more likely to have difficulties with everyday skills (mean difference = 5.2, 95%CI [0.7, 9.7]). There were no differences in the other domains including memory, orientation, abnormal behavior and motivation.

There was no statistical difference in the duration of symptoms and caregiving period between AD and

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Table 1. Demographic information of carers

	ALL (n = 33)	AD COHORT (n = 20)	BVFTD COHORT (n = 13)	P
Age at assessment	53.8 (SD 12.5)	53.6 (SD 11.7)	54.2 (SD 14.1)	0.907[m]
Sex	21 females (64%)	11 females (55%)	10 females (77%)	0.278[f]
Carer living with the patient	30 (91%)	19 (95%)	11 (85%)	0.547[f]
Living in a regional area	12 (36%)	6 (30%)	6 (46%)	0.465[f]

f = Fisher's exact test, m = Mann-Whitney U test.

Table 2. Demographic information of patients

	ALL (n = 33)	AD COHORT (n = 20)	BVFTD COHORT (n = 13)	P
Age at assessment	57.2 (SD 7.4)	58.1 (SD 4.8)	55.8 (SD 10.4)	0.725[m]
Sex	11 females (33%)	6 females (30%)	5 females (38%)	0.714[f]
Psychiatric history	13 (39%)	8 (40%)	5 (38%)	0.930[f]
Family history of dementia	12 (36%)	8 (67%)	4 (31%)	0.719[f]

f = Fisher's exact test, m = Mann-Whitney U test.

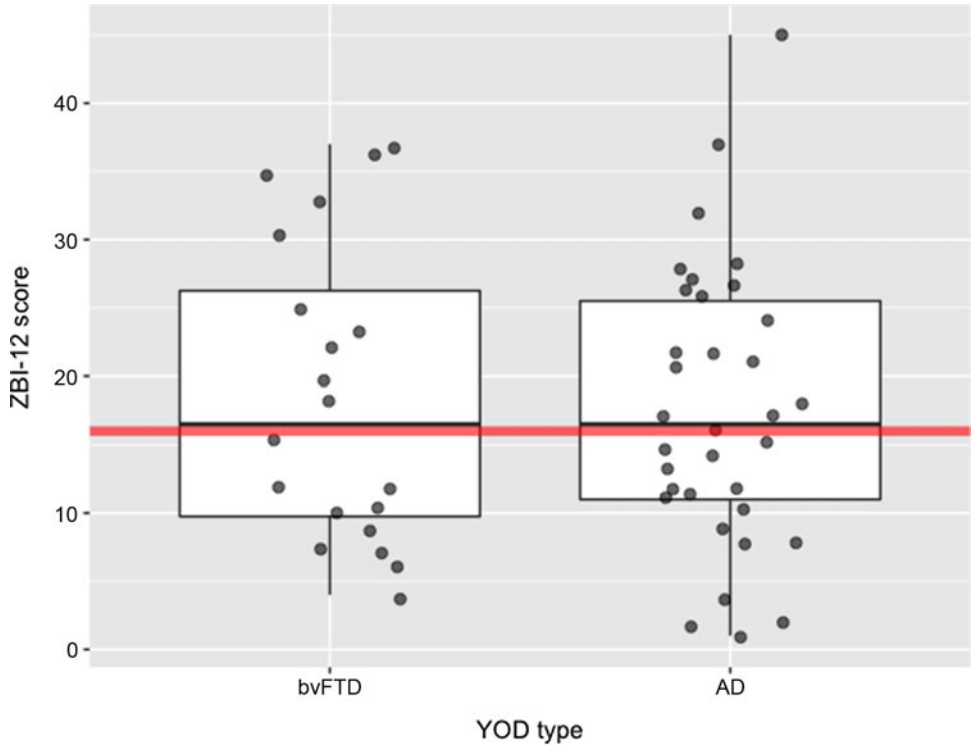

Figure 1. Boxplot of carer burden in YOD subtypes.

bvFTD as reported by the carers. The mean total NUCOG score overall was 63.6 (SD = 16.7), indicating moderate cognitive impairment, and was significantly lower in the AD cohort (mean difference = 14.0, 95%CI: [3.2, 24.9]). Similarly, those with AD had significantly lower scores in the NUCOG subdomains of attention, visuoconstruction, memory and executive functioning. Of note, five patients (all of whom were diagnosed with AD) had their NUCOG total score calculated from their MMSE score as they could not complete the NUCOG, with details provided in Table 4.

We found a significant difference in ZBI-12 scores between AD and bvFTD with total NUCOG scores as a covariate ($F = 11.11$, adjusted $R^2 = 0.387$, $p < 0.001$), as well as total CBI-R scores as a covariate ($F = 6.56$, $R^2 = 0.258$, $p = 0.004$). This should be interpreted with caution given the small sample size.

On comparison of CBI-R and ZBI, we found mood ($r = 0.642$, $p < 0.001$) and beliefs ($r = 0.532$,

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Table 3. Carer burden and clinical variables

	AD COHORT (n = 20) MEAN (SD)	BVFTD COHORT (n = 13) MEAN (SD)	MEAN DIFFERENCE [95% CONFIDENCE INTERVAL]	P^M
Carer burden	18.5 (14.0-22.9)	15.2 (10.5-21.0)	3.3 [-4.4, 11.0]	0.372
Duration of dementia symptoms	3.6 (2.7-4.6)	4.5 (3.0-6.5)	0.9 [-2.7, 0.8]	0.434
Duration of caregiving	2.4 (1.7-3.2)	3.6 (1.7-5.5)	1.2 [-3.8, 0.9]	0.682
CBI-Revised				
CBI total	67.4 (53.8-81.0)	63.3 (46.4-80.2)	4.9 [-15.7, 22.6]	0.624
CBI memory and orientation	20 (17.1-23.1)	15.8 (12.1-19.4)	4.3 [-0.6, 8.4]	0.800
CBI everyday skills	**11.4 (8.5-14.2)**	**6.2 (2.7-9.7)**	**5.2 [1.1, 9.0]**	**0.024**
CBI self-care	4.2 (2.0-6.3)	2.6 (0-5.3)	1.6 [-1.7, 4.7]	0.316
CBI abnormal behaviour	6.1 (3.4-8.8)	7.6 (4.3-10.9)	1.5 [-5.4, 2.1]	0.524
CBI mood	5.2 (3.6-6.8)	5.2 (3.2-7.1)	0 [-2.8, 2.9]	0.730
CBI beliefs	0.9 (0.2-1.5)	0.2 (0-1.0)	0.7 [-0.1, 1.5]	0.456
CBI eating habits	4.0 (2.1-6.0)	4.7 (2.3-7.1)	0.7 [-3.9, 2.3]	0.650
CBI sleep	2.7 (1.7-3.7)	1.6 (0.4-2.8)	1.1 [-0.4, 2.5]	0.181
CBI stereotypic and motor behaviours	**3.9 (1.9-5.8)**	**7.8 (5.4-10.1)**	**– 3.9 [-6.9, -0.6]**	**0.010**
CBI motivation	9.1 (6.0-12.2)	11.7 (7.9-15.5)	2.6 [-7.5, 2.2]	0.265
NUCOG				
NUCOG total*	**55.7 (43.4-60.1)**	**72.3 (65.4-79.1)**	**– 16.6 [-31.6, -9.9]**	**0.002**
NUCOG attention	**9.6 (7.3-11.8)**	**14.8 (12.2-17.4)**	**– 5.2 [-8.7, -2.8]**	**0.001**
NUCOG visuoconstruction	**11.0 (8.9–13.1)**	**15.8 (13.3-18.2)**	**– 4.8 [-7.9, -2.6]**	**0.002**
NUCOG memory	**8.4 (6.5-10.3)**	**12.0 (9.8-14.3)**	**– 3.6 [-6.7, -1.6]**	**0.005**
NUCOG executive	**8.9 (6.7-11.1)**	**12.4 (9.9-15.0)**	**– 3.5 [-6.8, -0.9]**	**0.027**
NUCOG language	14.0 (11.8-16.3)	17.0 (14.4-19.6)	– 3.0 [-6.8, 0.7]	0.057

CBI-R = Cambridge Behavioural Inventory Revised.
m = Mann-Whitney U test.
NUCOG = Neuropsychiatry Unit Cognitive Assessment tool.
ZBI-12 = Zarit Burden Inventory-short version.
Bolded = $p < 0.05$.
*Note: for cases where the calculated NUCOG total score was very low (ie <20 including 0), we assigned their score as 20.0 after discussion with experts including the original NUCOG authors. This was to avoid over-estimating their cognitive impairment which may have affected the statistical analysis.

Table 4. Details of participants unable to complete NUCOG

PARTICIPANT ID	DIAGNOSIS	MMSE TOTAL SCORE	CALCULATED NUCOG TOTAL SCORE	CLINICIAN COMMENT LACK OF NUCOG
3	AD	13/30	29.4	Patient was non-verbal.
31	AD	15/30	37.4	Patient with severe Alzheimer's disease with language and parietal difficulties;
39	AD	0/30	20.0*	Patient was disoriented and unable to do three words with very poor short-term memory.
41	AD	8/30	20.0*	NUCOG abandoned due to poor single-word retrieval
47	AD	15/30	37.4	Patient unable to complete NUCOG due to poor STM, with neuropsychology moderate dementia severity.

AD = Alzheimer's disease, NUCOG = Neuropsychiatry Unit Cognitive Assessment tool.
*Note: for cases where the calculated NUCOG total score was very low (ie <20 including 0), we assigned their score as 20.0 after discussion with experts including the original NUCOG authors. This was to avoid over-estimating their cognitive impairment which may have affected the statistical analysis.

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$p = 0.001$) domains to have a positive association with carer burden. Moreover, the total CBI-R score ($r = 0.541$, $p = 0.001$) was also significantly associated with carer burden. The other domains did not show significant correlation.

Discussion

Levels of carer burden were found to be high in younger-onset AD and bvFTD. However, we found no differences in burden found between the groups, in contrast to previous findings (Mioshi *et al.*, 2013; Uflacker *et al.*, 2016; Wong *et al.*, 2012). We also found high levels of behavior and motivation disturbance in both younger-onset AD and bvFTD groups. This contrasts to previous literature that found younger-onset bvFTD had higher levels in these behavioral domains compared to younger-onset AD (Mioshi *et al.*, 2013). Behavioral and emotional disturbance in general has been linked with increased carer burden (de Vugt *et al.*, 2006; Liu *et al.*, 2017; Wong and Wallhagen, 2014), but we failed to find differences in these behavioral disturbances between the two dementia subtypes which was unexpected (Wear *et al.*, 2008). This may explain why similar levels of carer burden were reported by the carers of the people with younger-onset AD and bvFTD.

We did find that carer burden was significantly different between younger-onset AD and bvFTD with total CBI-R score as a covariate, consistent with the literature that neuropsychiatric symptoms are associated with increased burden. We similarly found that carer burden significantly differed between the dementia subtypes is controlling for total neurocognitive scores, a marker of dementia severity. However, these findings should be interpreted with caution given the small sample size and the mixed findings in the literature as to whether cognition influences carer burden (Kang *et al.*, 2022; Kimura *et al.*, 2021; Miller *et al.*, 2013).

We also found that overall behavioral disturbance as well as mood and belief related were significantly associated with carer burden in YOD. This highlights the importance of clinicians considering all neuropsychiatric symptoms no matter the dementia subtype, given behavioral symptoms such as apathy and disinhibition are not specific to a dementia subtype, but are present across the dementia spectrum (Jenkins *et al.*, 2022). This is in line with the transdiagnostic approach promoted in recent literature (Cuthbert, 2022; Jenkins *et al.*, 2022).

Our findings highlight the importance of clinicians routinely screening for behavioral disturbance in YOD, irrespective of presumed pathological diagnosis. Moreover, these observations are concordant with observations that YOD may present with psychiatric symptoms initially (Ducharme *et al.*, 2020; Tsoukra *et al.*, 2021; Woolley *et al.*, 2011) and early referral to a specialist YOD service might facilitate a more seamless assessment and timely diagnosis for dementia (Loi *et al.*, 2022). The routine use of standardized measures such as CBI-R or Neuropsychiatric Inventory (NPI) likely assists clinical discovery where carers might otherwise be reluctant to disclose these if their loved one with dementia is present during the review.

Firm conclusions about the lack of difference in carer burden are prevented by the small sample size and limited power of the study. The authors attempted to mitigate the limitations of the sample size by using robust statistical methods to reduce variance. Further, we were unable to investigate the effects of other individual factors on carer burden such as YOD subtype, type or severity of behavioral disturbance and carer-related information such as relationship, sex or co-residing was related to carer burden. We recommend future studies to incorporate larger sample sizes to better characterize the multifaceted causes of carer burden. The retrospective study design also has limitations regarding reporting bias and case notes. Furthermore, results may not be generalizable to participants who are not fluent in written English, restricting extending these findings to other cultural and linguistic diverse populations. Additional measures such as identifying additional neuropsychiatric symptoms, staging of dementia and severity of dementia symptoms may also help clarify other relationships. Finally, this is a single-site study at a tertiary service with most participants from one state, which affects our findings' generalizability and external validity.

In conclusion, this study reports on the high levels of behavioral disturbance and burden experienced by carers of younger-onset AD and bvFTD. These findings highlight that despite the younger age, it is important for clinicians to screen for behavioral disturbance in all people with YOD regardless of subtype. Further study might involve a more detailed examination of the factors associated with carer burden so that appropriate interventions to target burden can be developed.

Conflict of interest

All the authors report no conflicts of interest related to this work.

Description of authors' roles

MK: Study design, data collection, statistical analysis, manuscript production and editing.

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SF: Study design, data collection, manuscript production and editing. AE: Study design, manuscript editing, WK: Study design, data collection, manuscript editing, MW: Study design, statistical analysis, manuscript editing. DV: Study design, statistical analysis, manuscript editing. WHC: Data collection.

DE: Data collection, study design, manuscript review and editing. SL: Study design, data collection, statistical analysis, manuscript review and editing.

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Ethics approval statement

This study was approved by the Melbourne Health Research Ethics Committee (approval numbers 2016.038 and 2018.371).

Patient consent statement

Participants provided written consent unless there was a capacity issue in which case their substitute decision-maker (next of kin) consented on their behalf.

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