

HUNTINGTON'S DISEASE AND RESIDENTIAL AGED CARE

Submission to the Aged Care Royal Commission

Endorsed by

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SUMMARY OF SUBMISSION

This submission primarily relates to the challenge of providing care to Australians with disabilities caused by Huntington's disease, living in residential aged care, particularly younger people with disabilities

Huntington's disease (HD) is a hereditary neurological disease that causes profound and relentless progressive physical, cognitive and emotional deterioration, which leads to very high levels of disability and premature death. At present, no cure and no effective treatments exist for HD. Eventually, most people with HD require residential care. For over 75% of people with HD in Australia, the only option is residential aged care even though most will be entering residential care while considerably younger than the threshold age of 65.

Personal stories and case studies in this submission give examples of the experiences of younger people with HD in the Australian residential aged care system (page 11).

Recommendation

The residential care needs of people with HD are well understood. For effective outcomes and the most efficient use of resources, we recommend funding and supporting providers via the NDIS who will supply a mix of residential care environments specifically designed for people with HD, in age-appropriate residential facilities, rather than the current approach of locating people with advanced HD in generalist aged care homes, often in locked dementia wards. We also recommend relevant training for any providers and their staff caring for people with HD.

WHAT IS HUNTINGTON'S DISEASE?

IMPACT

Huntington's disease (HD) is a hereditary neurological disease that causes progressive and relentless physical, cognitive and emotional deterioration. The most common age of onset is between 35 – 50 years although it can occur at earlier or later ages. The course of HD is usually between 15 – 20 years with the disease eventually causing premature death.

Huntington's disease is autosomal dominant and each child of a parent with HD has a 50% chance of inheriting the mutation in the gene and developing HD.

TREATMENT

No cure and no effective treatments for HD exist, but a highly co-ordinated global research effort is actively seeking ways to treat it, and professional caregivers are developing better ways to improve the quality of life of those affected, including through exercise and occupational and other therapy.

PREVALENCE

A study in NSW in 2018 found the prevalence of people with manifest HD to be 7.5 per 100,000 of the population.¹ Extrapolating across Australia's population of 25.54 million², the number of people with HD is about 1,920. However, the Tasmanian situation is worse, with a prevalence in excess of 20 per 100,000. Given the hereditary nature of HD, this means another approximately 8,000 people are at risk.

However, we recognise that the NSW study is now considered conservative. Several international studies have identified prevalence of up to 12 per 100,000, and anecdotal evidence from some of our state associations points to an even higher prevalence as new families become known to them, indicating a potential prevalence in excess of 3,000.

DO PEOPLE WITH HD NEED RESIDENTIAL CARE?

HD is characterised by uncontrollable movements, cognitive and emotional impairment, behavioural issues, and speech and swallowing difficulties. 'The behavioural changes are often the most distressing part of the condition and create the greatest challenge for the person with the disease, their family and professional carers.'³

As the disease and associated disability progresses, the person requires increasingly higher levels of care. Such care is usually too onerous for the person caring for them at home, especially considering that this need will continue to increase for many years. Eventually, for most people, residential care of some kind becomes the only option. Typically, the age at which someone with HD enters residential care will be between 40 and 60.

Due to frequent breakdowns in family relationships because of the nature of HD, a number of people live alone and require community-based support services from an early stage of the disease. Because of cognitive dysfunction and lack of insight, they may refuse such services, thus placing themselves in dangerous and unhygienic situations (for example, spilling boiling water, not showering or changing their clothes, and so on). Thus, a move to residential aged care may be the only available solution to these difficult circumstances.

In some instances, group homes have been tried with varying degrees of success, depending on the type of support available to residents. Failure of the arrangement is usually due to the cognitive impairment and behavioural issues associated with HD. However, while a group home or similar community residential arrangement (with appropriate support services) may meet the

needs of some, eventually these residents will still need to move to residential care of the type currently supplied by aged care homes as their HD progresses.

The Huntington's Outreach Service (Westmead Hospital, Sydney) estimates that among those under the age of 50 in residential aged care in NSW, about 10% have HD.

We note that people with HD are often forced to move around to different aged care homes due to inability to provide adequate care, and/or be admitted to hospital inappropriately, usually as a result of perceived behavioural issues. This is both disruptive and upsetting to the person with HD (and their family), as well as being expensive and time-consuming for health/service providers.

CURRENT SITUATION WITH RESIDENTIAL CARE

RESIDENTIAL AGED CARE

For most people with HD, the only option when residential care becomes necessary is to move to an aged care home. A small number of people are able to access some specialised facilities in NSW, Victoria, SA and WA.

Table 1.

Younger people living in aged care 30 September 2018 (Source: Dept of Health)	
Under 65	5,905
Under 55	1,179
Under 45	188
Under 35	30

Table 2.

	30/06/14	30/06/15	30/06/16	30/06/17	30/06/18
People <65 with Huntington's disease admitted to residential aged care last 12 months	59	49	58	41	47
People >65 with Huntington's disease admitted to residential aged care last 12 months	30	41	43	35	40
Permanent residents <65 in aged care with Huntington's disease	167	179	185	172	162
Permanent residents >65 in aged care with Huntington's disease	165	179	185	192	186

Source: personal communication from AIHW 2019

The data show that close to 60% of people with HD in residential aged care are under 65 years of age. Furthermore, we believe most people with HD entered residential aged care when they were under 65.

OTHER FORMS OF RESIDENTIAL CARE AND HOUSING FOR PEOPLE WITH HD

Specialised HD units

Three states (NSW, Victoria and WA) have some places in residential facilities that specialise in the care of people with HD - a total of 62 places, with significantly higher staffing ratios that are typically found in residential aged care:

Table 3.

Jurisdiction	Specialist facility	Places	Provider
NSW	Huntington's disease Unit, St Joseph's Hospital, Auburn	14	St Vincent's Hospital Network
VIC	Arthur Preston Residential Centre, East Burwood	30	Wesley Mission Victoria (no longer exclusively HD)
WA	Kailis House, Belmont (early stage)	6	Brightwater Care Group
WA	Ellison House, Carlisle (late stage)	12	Brightwater Care Group

Other ways people with HD may be accommodated

Examples of other housing options:

- Three people in Queensland, nine people in Tasmania and eight people in South Australia receive 24 hour care in community housing. At least 20 people in NSW are also known to the Association to be receiving 24 hour care in the community (including NDIS Supported Independent Living (SIL)).
- In WA some people with HD find accommodation and care in other specialised facilities such as: Cerebral Palsy Shared living Home (metro); MS Treendale Gardens, young disabled care residential facility (country); and MS Hamilton Hill.
- A person with advanced HD has recently moved from a secure dementia unit in an aged care home to a specialist disability accommodation (SDA) home with 4 beds in regional NSW,

with care funded by from the NDIS SIL. She is much happier, has put on weight and has a staffing ratio of 1:4 as opposed to 1:19 in the aged care home.

WHAT'S WRONG WITH RESIDENTIAL AGED CARE FOR THOSE WITH HD?

- We note that there have been a number of federal and state government inquiries into the issue of younger people living in aged care homes (e.g. Senate Inquiries 2004, 2015). Without exception they have produced nothing of value for this cohort.
- The average age of all residents in residential aged care has been increasing. At 30 June 2018, 65% of permanent residents were aged 85 and over, up from 57% in 2011, and 50% in 2000. Stays in residential care are becoming shorter and residents have a more complex array of health conditions.

Against this background, it can be quite devastating for a younger person with HD to move into an aged care home for what is usually a stay of many years. It is true that HD can affect a person by making them more self-centred and less aware of their surroundings. Even taking this into account, family and friends can find themselves visiting their loved one in very challenging circumstances.

- For younger residents with HD, aged care facilities lack access to age-appropriate social activities and interaction with the residents' peer group. Ideally, younger residents should have access to an enriched residential environment that gives them some reasonable quality of life. In fact, some recent research is suggesting that an enhanced environment can marginally ease some symptoms of HD¹. Instead, younger people with HD are mostly housed with and treated the same as the very elderly.
- A lack of specialist ongoing training available for professional care givers combined with a general lack of adequate staffing means that the nature and quality of care can be expected to be inadequate. Indeed family carers are almost universal in their condemnation of their relatives' care experiences and outcomes.
- A frequent example of the lack of appropriate care outcomes is the clinical need for people with Huntington's disease to maintain high calorific intake due to their chorea. Residents are reported to be effectively starving in residential aged care due to being left to feed themselves at meal time, then sent back to their rooms when time is up despite having eaten little.
- People with HD in aged care facilities are distributed over many individual facilities and across all regions of the country so that, without a clear pathway of specialised nursing and care training, the quality of care delivered is quite variable, despite the best intentions of the institutions engaged in delivering care.
- Some people may be able to stay at home longer if respite care and appropriate in-home support services were more readily available.
- Furniture: there is often a need for HD-specific beds and chairs to avoid potentially significant injury to the person with HD, and carers. In our experience few aged care facilities are able or willing to provide this expensive specialised furniture.

¹ Cruickshank TM, Reyes AP, Penailillo LE, et al. Effects of multidisciplinary therapy on physical function in Huntington's disease. *Acta Neurol Scand.* 2018; 00:1-8. <https://doi.org/10.1111/ane.13002>

WHAT WILL BE THE IMPACT OF THE NDIS?

- An objective of the NDIS is to assist younger people currently in residential aged care to find 'community-based settings or other age-appropriate settings where possible.'⁴—that is, to move back into the community. However, the reality is that HD is degenerative: a move back into the community is highly unlikely for a person with HD. It is when all other avenues of care have been tried and they can no longer live in the community, even with a range of support services, that the person with HD requires residential care.

Therefore, whatever changes the NDIS brings, they must take account of the fact that people with HD will ultimately require residential care with a high level of support services in their final years — they will not be returning to the community.

- As the data show (see Table 2), many people with HD receive residential care in the aged care system; some others receive care through specialised units or community-based housing with appropriate levels of 24-hour care. Given the lack of a uniform approach nationally, and the different sources of funding for support services, it is likely that the introduction of the NDIS will have varying impacts on people depending on their specific circumstances. However, the few specialised HD units around the country point to what is needed to provide best practice care for HD residents. The key to a successful move to the NDIS will be to fund for this best practice level of care.
- Those people with HD being cared for in community housing will need funding from the NDIS at a level adequate to provide the necessary support services that enable them to stay in that kind of accommodation—particularly those services that compensate for the impact of cognitive impairment and the behavioural issues associated with HD. However, we need to be realistic and recognise those residents will ultimately need to go into full residential care.
- Due to the cognitive changes associated with HD, it is essential that NDIS-funded Service Assessors, Local Area Coordinators, Planners, Support Coordinators and registered providers are familiar with HD, recognise the reason support is needed and the impact of degenerative cognitive impairment.

HOW TO ADDRESS THE RESIDENTIAL CARE NEEDS OF PEOPLE WITH HD

Because HD is a hereditary disease with a well understood progression, its impact on the demand for residential care can be forecast, within reasonably predictable limits. This removes a large measure of uncertainty from the costing of public policies relating to the care of those with HD. Therefore, a rational and holistic approach to dealing with the care needs of those with HD is to fund directly residential care through providers who are able to provide both suitable physical facilities and trained nursing and care staff - that is, fund providers who will supply residential care specifically designed for people with HD rather than the current approach of caring for people with HD as if they were frail elderly people with dementia.

The process to reach this point **recommended** by this submission:

1. Identifying and costing the best practice model(s) of care for residential care of people with HD.

2. Providing funding for training professional carers (nurses, allied health professionals and carers).
3. Approving and funding residential care providers via NDIS who can provide suitable physical facilities and trained professional staff. Places need to be funded, according to need, in both metropolitan and regional areas via Specialist Disability Accommodation to cover disability specific building requirements.
4. Funding to participants with a care model for Supported Independent Living (SIL) in Huntington's specific residential care.

This important shift in support arrangements would mean that people with Huntington's would no longer be discriminated against in the aged care system, regardless of their age, as they lose out if under 65 and again in different ways if over 65.

PERSONAL STORIES AND CASE STUDIES

A PERSONAL STORY

I am currently caring for my 59-year-old husband who has Huntington's disease. As this is a genetic illness, we have had the misfortune of visiting other family members who have been either in respite or full-time care in aged care facilities. Taking our young children to visit their 46-year-old uncle and their 60-year-old grandfather in a geriatric facility where some people in the same ward were in their final stages had such an adverse effect on our children we had to stop taking them to visit.

Now that my husband is having respite on occasion, it is totally unacceptable that he has to be placed where he has nobody with interests or people that he can relate to. Group activities and music are all geared for people nearly twice their age. It is incredibly depressing for the person in care as well as family or friends visiting.

I realise we are in the minority; however, young people who require care or respite should be afforded the same respect and concern and dignity that is given to the aged senior citizens. They have already given and lost so much. We were unable to have respite at one aged care facility as my husband wasn't eligible: he was too young. They only took people over the age of 65, even though he has been ACAT-assessed and has only ever been in aged care facilities. Where does that leave us, yet again in a hopeless situation? The powers that be need to understand and realise that there have to be hostels and nursing homes that can accommodate young people - the need is greater than you realise, and it needs to be done as soon as possible.

Sincerely,

DF, Central Coast NSW

A PERSONAL EXPERIENCE

How has placing a young family member in age care affected the young, you and your family?

What do you think is inadequate about having young people accommodated in aged care? Please provide some personal experiences.

Our experience relates to the placement of two brothers in the advanced stages of Huntington's disease, one aged 44 and the other in his mid-fifties at the time of placement. They were placed in nursing home age care in two separate facilities in the Newcastle region. The standard of accommodation, facilities and services (and unfortunately the skill of the staff, primarily because of the lack of knowledge of Huntington's disease and the management of HD sufferers) had and continues to have an effect on the frequency and length of visitations by close family members, especially in one case the children of the resident, who had feelings of helplessness and despair for the hopeless situation of the residents. All these issues impacted on the attitude of the residents, who regularly expressed fear and anxiety about being left by their loved ones in such a "horrible" place. Neither home provided suitable or appropriate activities for younger people. Even the sing-along sessions in one home did not cater for "younger" patients. Consequently, the songs not triggering any memories in them, the younger patients had no wish or interest to participate.

Standards in nursing homes vary from excellent to poor, depending on one's financial circumstances and the requirements of the resident (age care or otherwise), and facilities that suit the resident's needs are not always available close to the resident's family and friends. In regional areas this is even more problematic. In placing a loved one in care, when financial reality,

proximity and convenience win over “resident needs”, the resident’s attitude suffers, and this in turn, as the resident’s health and well-being deteriorate as a result, has a compounding effect on the attitude of family and friends.

Living with aged people, and being treated and managed as an aged person, impacts on a younger person’s attitude and self-esteem, and without activities and treatment processes that stimulate the mind, and in turn attitude, confidence is lost and positive feelings and thoughts are replaced by negativity and hopelessness.

Every effort should be made to ensure compliance with the United Nations Convention on the Rights of Persons with Disabilities, which states that all people with a disability should have the “opportunity to choose their residence and where and with whom they live on an equal basis with others, and not be obliged to live in particular living arrangements”.

Programs such as the National Disability Insurance Scheme should seek to minimise as much as possible the placement of young people in nursing home age care and concentrate on establishing opportunities for them to continue to live in the community by offering more options for age-specific, and even disease specific, group “home-style” accommodation, which would have an enormous beneficial impact on their feelings of independence and self-worth.

What do you think can be done to improve circumstances for aged care?

- Specialised Disability Accommodation for Huntington’s specific residential care.
- Separate wing or ward in nursing homes for younger patients to enable them to interact with one another.
- Additional and more regular age-appropriate activity programs.
- Higher standards of training for staff related to younger patients and their conditions; e.g. Huntington’s, Motor Neurone Disease, etc.
- Additional assistance to carers in their own homes to delay as long as possible the placement of loved ones in care.
- Higher standards for accreditation (and strictly enforced) for the accommodation and care requirements of younger people in care.
- Increase staff levels to allow staff more intimate, personal time with younger people to promote independence and confidence in their performance of everyday tasks, especially those with communication and comprehension difficulties. At the moment nursing homes cater for the elderly and younger people alike, with little differentiation in the management and appropriateness of care. They should not all be put in the one basket: care in nursing homes should be flexible, and as an extension, the types of services offered by staff should be flexible also to suit the individual needs of all patients.
- Consideration should be given to limiting the number of large nursing homes in favour of smaller group community homes for younger people - ideally specific homes that cater for specific disabilities and diseases - creating a home-style environment that allows patients to communicate with people in similar circumstances, and one that promotes a more independent lifestyle, rather than the time-constrained, institutional rigidity of nursing home care.

Contributors – M & D, Central Coast NSW

FEMALE CLIENT A, AGE 46

Located in Regional NSW, married with 2 children, aged 18 and 21.

Living in a rural setting, the family relied on home-based services to assist, but found that those services could not meet the significant needs of the family and, Client A in particular, as the illness progressed.

The family decided after several stays of 2-3 weeks in respite care, that permanent residential care was required for Client A. This decision was made in consultation with relevant health professionals. The choice of aged care home was quite limited if they were to keep Client A relatively close to home allowing easy visiting access for the family.

An issue faced when the possibility of an aged care home placement was raised was a concern with going into a care facility full of 'old people'. This concern was realised when Client A moved to the aged care home.

The initial move to permanent care lasted a few months. Client A was not happy there, and the family felt that the care provided was not to an acceptable standard. Concerns included an apparent lack of understanding of Client A's needs as a younger person, and of HD specific issues.

Client A returned home with provision of some services in home, but this did not work out as Client A deteriorated, and her husband—the main carer—could no longer manage, even with services to assist.

This resulted in seeking a position in a different aged care home. This has been more successful, but not trouble free.

Challenges faced in each aged care home have included providing activities of interest to Client A as many of her interests and abilities differ markedly from those of other residents. As well, staff have not treated Client A in line with her age and illness, given her situation is quite different from most residents.

MALE CLIENT B, AGE 56

Located in metropolitan WA, widowed with 2 sons

Client B was widowed in 2010. He remained at home with his 19-year-old son with support from Home and Community Care. An older son took over his care two years later. From 2013, it became more difficult to support Client B at home and a suitable placement was sought. It was 12 months before a placement, in an aged care facility, became available. The adjustment for Client B from home to living in an aged care facility was very difficult. He had little in common with other residents who were much older than him. In addition, the care environment was not conducive to either his grandchildren or other immediate family members visiting. Client B became isolated, withdrawn and very unhappy.

The situation changed for Client B when he was able to be placed at a residential care facility for younger people with a disability. In this facility, Client B has his own room and a small living area with a kitchenette. Since being there his family has re-commenced visiting on a regular basis. Client B feels very comfortable having his grandchildren come to his room. It gives privacy and they can visit without needing to engage with other residents in the facility. Client B reports he is much happier now that he is not living in aged care. This placement is only possible as Client B was successful in obtaining CAP funding from the Disability Services Commission (WA).

MALE CLIENT C, AGE 40

Located in Regional NSW, married with a child aged 9.

Client C was living at home but fairly rapid deterioration physically, cognitively and behaviourally necessitated placement in an aged care home when he was aged 38.

There was an initial perception issue for Client C with the concept of going into an aged care home full of 'old people'. Once the need for care was identified, there was a real issue finding an aged care home in reasonable proximity to home that would accept younger people. Client C was initially placed in an aged care home 100km away from home, as no place could be found locally. A placement closer to home became available about two months later.

Client C reacted very badly to some nursing staff, possibly due to their lack of understanding of HD symptoms, and his reaction to some staff of non-Australian background was very aggressive.

This challenging behaviour resulted in the aged care home seeking expert assistance which led Client C being placed in the local hospital, then moving to a short-term acute care facility for several months. At this time, further training was provided to the aged care home, and the Dementia / Behaviour Assessment and Management Service was engaged to assist. Client C subsequently returned to the aged care home, and the situation is significantly improved.

Client C's wife and child have found it very difficult visiting him in the aged care home setting where he is so much younger than other residents.

FEMALE CLIENT D, AGE 43

Located in metropolitan WA, single

Client D is single and prior to her admission to a low-level aged care facility was living independently. She was supported in her independent living by her older, unwell mother. Prior to her first admission to an aged care facility, she had several hospital admissions for acute mental health issues which were followed by extended periods of emergency respite. Client D was admitted to low level aged care in 2008 at age 41. Client D did not settle in aged care and was discontented and unhappy. She remained in aged care until 2014 when her mother was eventually able to secure a place in a group home for younger people. This option became possible as a result of a funding package through the state Disability Services Commission (WA).

A TASMANIAN EXAMPLE

Re-located from Tasmania to Victoria

A younger person with HD in Tasmania (now 40 years of age) spent two years in aged secure Mental Health Service care—their long-term accommodation unresolved. The permanent solution found has been to move them to accommodation in Victoria.

¹ McCusker, Elizabeth A., Reynolds F. Casse, Shanthi J. Graham, David B. Williams and Ross Lazarus, *Prevalence of Huntington disease in New South Wales in 1996*. Medical Journal of Australia, 173 (August 2000), 187-190. This study by McCusker et. al. is considered the best study on prevalence in Australia. A follow-up study by Dr Clement Loy (current Director of the Westmead HD Service) to be published later this year is expected to find an incidence of 7.5 per 100,000. We note that data from Tasmania indicates a much higher prevalence there of 12.1 per 100,000 (due to a founder effect), which should be taken into account when developing future public policy.

² Australian Bureau of Statistics, Population Clock, <http://www.abs.gov.au/ausstats/abs%640.nsf/94713ad445ff1425ca25682000192af2/1647509ef7e25faaca2568a900154b63?OpenDocument> accessed 4 December 2019

³ Snowden, Dr Julie, *Understanding Challenging Behaviour in Huntington's disease*, 14th Meeting of the International Huntington Association, Denmark, August 2001.

⁴ *Younger people in residential aged care: NDIS eligibility meeting*. Information sheet published by the National Disability Insurance Scheme (updated 22/11/17). Available for download at <https://agedcare.health.gov.au/programs/younger-people-in-aged-care/younger-people-in-residential-aged-care-ndis-eligibly-meeting>