Living well with Huntington's Disease: Community of Practice Framework

A GUIDE FOR PRACTITIONERS AND PROVIDERS



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Forward - Dr Patricia Kailis AM OBE

I have been involved with families living with Huntington's Disease for over 25 years, and it is inspiring and inspirational to work alongside these special people.

Huntington's Disease is a complex condition that alters lives and challenges family dynamics and yet we continually see extraordinary resilience and hope among the families who deal with it on a daily basis.

More than anything, seeing families cope with these extraordinary circumstances strengthens our resolve to ensure that they have the best possible services available to support them throughout their lives and their journey with Huntington's.

The desire to ensure this was the catalyst from which this framework was borne - to paint a vision for how things should be, and to take a holistic and collaborative approach to providing support which is based on evidence-based best practice.

This framework is a first for Western Australia and many people have been involved in its evolution. The lead organisations - Huntington's WA, the NMHS Neurosciences Unit, Brightwater Care Group, Department of Communities (formerly Disability Services Commission), MSWA and South West Huntington's Network - have provided invaluable oversight and experience that has ensured the community of practice and other agencies work together to provide high-quality care into the future.

The vision is to provide services for people living with Huntington's which are person-centred, holistic, and focussed on wellness, hope and resilience. The framework provides an outline for supporting people throughout their lives with collaboration, respect and above all dignity.

Dr Patricia Kailis AM OBE
Patron, Huntington's WA (Inc.)

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Purpose of the framework

The Community of Practice Framework embeds principles of care that support the lifelong enablement of people living with Huntington's Disease in Western Australia. The framework provides a clear vision for the management of Huntington's Disease, while guiding health practitioners and service providers to support and empower individuals with Huntington's Disease and their families to engage actively in their care and increase control over their health and wellbeing.

It adopts a lifespan approach, aiming to facilitate optimal health and wellbeing from early childhood to the end of life, irrespective of a person's age, severity of impairment, or place of residence. To achieve its purpose, the framework provides a set of care principles that reflect current evidence and contemporary practice. This includes an overarching focus on wellness and enablement, building on an individual's strengths, capacity and personalised goals to help them to achieve maximal independence, safety and participation at all stages of their journey with Huntington's Disease.

In accordance with the Ottawa Charter, the framework views health as a positive concept emphasising social and personal resources, as well as physical capacities. This considers the interaction between biological, psychological, social and environmental factors as important determinants of health and wellbeing.

More specifically, the framework aims to:

- Empower and enable people with Huntington's Disease to be active partners in care
- Guide proactive, ethical, and reflective practice
- Enable health practitioners and services providers to embed a wellness and enablement approach into every client contact
- Promote a lifespan and family approach
- Facilitate access to early intervention and support
- Promote Huntington's Disease friendly environments
- Facilitate effective collaboration and care integration across agencies and services
- Improve quality of care and guide service evaluation and quality assurance
- Assist people with Huntington's Disease and their families to understand their rights to ensure principles of care are upheld

The framework is built on understanding of the complex challenges associated with the disease and the barriers to good health faced by individuals living with Huntington's Disease and their families. It aims to optimise health and wellbeing within the recognised constraints of the condition. The framework articulates a practice approach for Huntington's Disease services that puts the individual living with Huntington's Disease at the centre, is family led and culturally responsive.

Background

Huntington's Disease is an incurable, life-limiting adult-onset hereditary neurodegenerative disease characterised by a triad of symptoms, including cognitive disturbance, motor abnormalities and psychiatric features (Paulsen, Hoth, & Stierman, 2005). Huntington's Disease affects both men and woman and results in the gradual loss of ability to carry out everyday tasks, with individuals becoming increasingly dependent on others over the course of the disease (Keenan et al., 2013).

The age of onset of Huntington's Disease is highly variable, with an average of approximately 45 years of age (Ross et al., 2014). As such, symptoms typically present during a significant life-phase in regards to childbearing, child-rearing and career development (Vamos et al., 2007). The age of onset and rate of progression of Huntington's Disease are influenced by both environmental and genetic factors (Ross et al., 2014).

The course of Huntington's Disease can be divided into pre-symptomatic, prodromal and symptomatic periods. In the pre-symptomatic stage individuals have no symptoms and are indistinguishable from healthy controls. A prodromal period may be detected, characterised by subtle motor, cognitive and behavioural changes, with minimal functional impact. Once motor and cognitive signs and symptoms emerge, they progress relentlessly over the course of the disease, which is uniformly fatal (Ross et al., 2014). Life expectancy is approximately 15 to 20 years following the onset of symptoms (Keenan et al., 2013).

Since 1993 a genetic test has been available to inform healthy individuals whether or not they carry the mutated gene which causes Huntington's Disease. An individual's decision to undertake genetic testing is highly variable, personal and complex. There are currently no disease-modifying treatments available, representing a priority in Huntington's Disease research (Ross et al., 2014).

Huntington's Disease should be conceptualised as a life-long and family condition. As an autosomal dominant disease, any child of a parent with Huntington's Disease has a 50 per cent chance of inheriting the condition. As a result, children witness the profound physical, cognitive and psychological decline of a parent, with the knowledge that they are at risk of the disease. Furthermore, children are often required to assume caregiver roles at a young age, extending the psychosocial impact of Huntington's Disease far beyond the individual themselves. The inter- and intra-generational impacts of genetic disorders are understood (Bombard et al., 2011) and Huntington's Disease is recognised as one of the most devastating illnesses because of its progressive nature and impact on families (Klimek et al., 1997).

Individuals living with Huntington's Disease and their families retain the same rights as anyone else; however, the nature of the condition means that people with Huntington's Disease are vulnerable members of society. Huntington's Disease has always been a

particular target for social stigma and discrimination (Bombard et al., 2011). Bombard et al. (2011) found that fear of genetic discrimination is nearly universal among individuals at risk of Huntington's Disease, relating to insurance, family and social settings; and to a lesser extent employment and health care settings. Studies suggest elevated rates of suicidal ideation in people at risk of and diagnosed with Huntington's Disease (Paulsen et al., 2005).

Individuals living with progressive neurological disorders like Huntington's Disease express a desire to remain in their homes and active in their communities for as long as possible (Giles & Lewin, 2008). Experts in the field support the notion that people with Huntington's Disease benefit from living in specialised, supported accommodation facilities where their extensive care needs can be met (Dawson et al., 2004). People with Huntington's Disease have complex personal care needs that require timely, appropriate and increasing levels of care as the condition progresses over time (Giles & Lewin, 2008). In the advanced stages of the disease, residential aged care is often the only care option (Dawson et al., 2004). Mainstream aged care placement, however, is often inappropriate given the relative youth of most people affected by Huntington's Disease (Dawson et al., 2004).

Research has demonstrated the complexity of the needs for specialist service provision for individuals with Huntington's Disease, their families and caregivers (Dawson et al., 2004). Coordinated efforts are required to ensure the rights of people with the disease and their families are protected. This includes young people living in Huntington's Disease families who may be at risk of developing the condition.

Family members often care for individuals with Huntington's Disease until a point where physical, cognitive and psychological symptoms exceed capacity for home-based care (Williams et al., 2008). This requires adoption of caregiving roles and tasks that can result in positive experiences, but may also involve burden or challenges for physical and mental health (Williams et al., 2008). Research indicates that carers of people with Huntington's Disease typically deal with problems as they arise, rather than adopting a more strategic approach to care (Williams et al., 2008). Williams et al. (2008) found that carers face emotional barriers and conflicting demands on time and resources that limit their ability to look beyond present challenges. Such research provides advice regarding how carers can and should be supported to help anticipate and prevent crises from occurring.

Management of Huntington's Disease in Western Australia: Looking across the continuum of care & Huntington's Disease trajectory

The management of Huntington's Disease in WA is coordinated by key agencies that form the core membership of the Huntington's Disease Community of Practice (see Page 9).

Huntington's WA (HWA) is a non-profit organisation formed specifically to improve the quality of life for people with Huntington's Disease, their families and carers in Western Australia. The association is dedicated to providing individual advocacy and support for the Huntington's community. The four major areas of support include community development, counselling and individual support, information and education, and research.

The Neurosciences Unit (NSU) is administered by North Metropolitan Health Service, Mental Health, a statewide service that provides specialised diagnostic support to both private and public hospitals and community-based services. The unit provides extensive services to families affected by Huntington's Disease. The multi-disciplinary team consists of clinical neuropsychologists, psychologists, social workers and speech pathologists. The unit also has a consultant neurologist and consultant psychiatrist specialising in neurodegenerative disorders. Huntington's Disease services include predictive testing and ongoing welfare management.

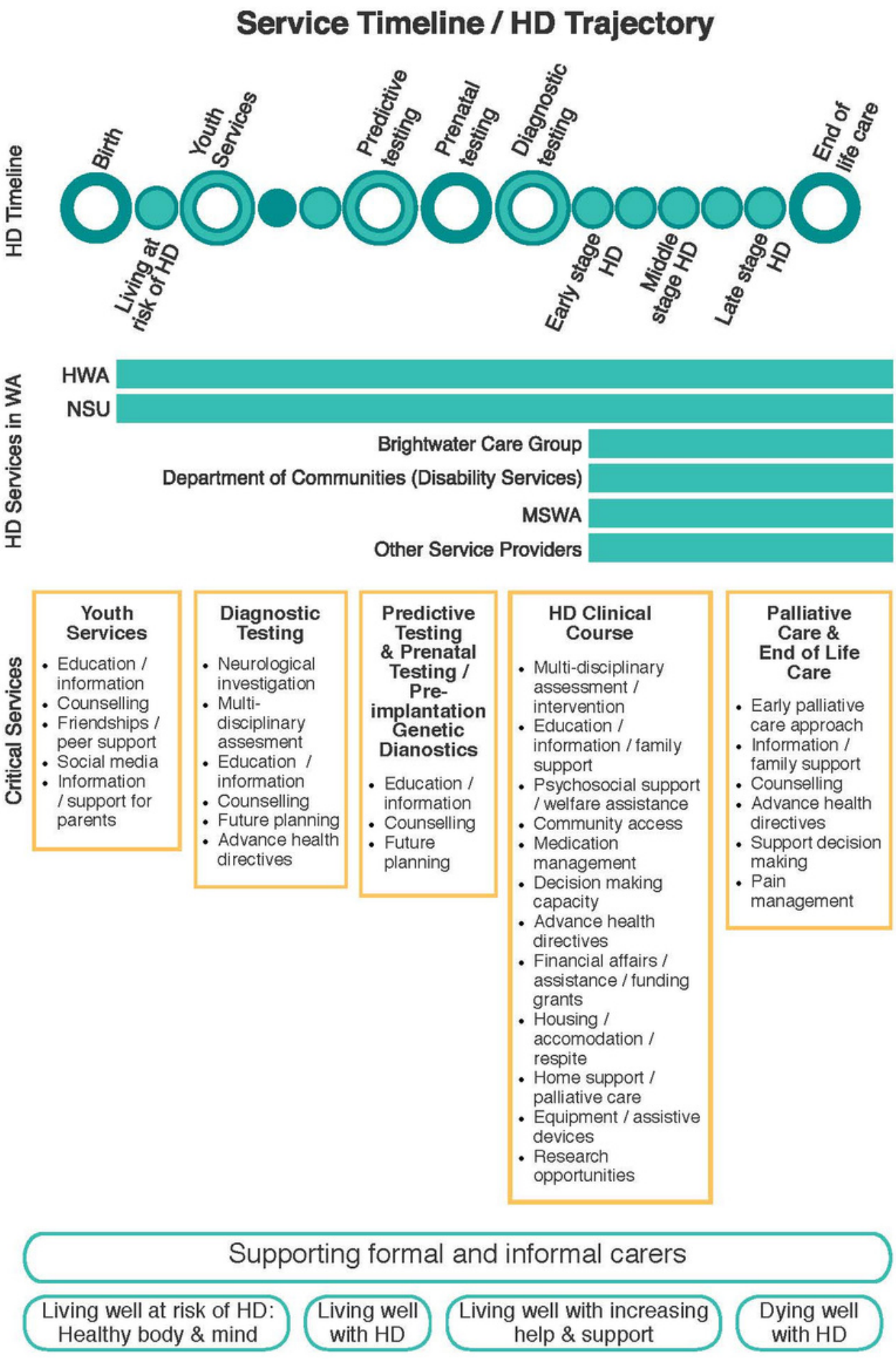
Brightwater Care Group provides accommodation services across a range of housing options in the Perth Metropolitan area, equipped to provide complex physical, cognitive and behavioural support. Brightwater operates two residential services for people living with Huntington's Disease, Kailis House and Ellison House. Both homes provide a supportive environment designed to promote independence, personal choice and lifestyle, while meeting the complex care needs of people living with Huntington's Disease.

Interagency meetings support coordination of services for individuals with Huntington's Disease and their families across the continuum of care. Feedback mechanisms between HWA and NSU (e.g. monthly meetings, client-by-client basis) further facilitate integrated care while both agencies are providing services. Figure 1 highlights the points along the trajectory of Huntington's Disease where each agency is most actively involved. The Community of Practice Framework was formed to enhance collaborative working across agencies, to improve standards of care and ultimately to optimise the health and wellbeing of individuals living with Huntington's Disease, their families and carers in WA.

Members of the Huntington's Disease Community of Practice Framework

Members of the Community of Practice Framework
Huntington's WA (HWA)
Neurosciences Unit (NSU), North Metropolitan Health Service, Mental Health
Brightwater Care Group
Department of Communities (Disability Services)
MSWA
South West Huntington's Network
Other agencies providing care to people with Huntington's Disease in WA
Acute, sub-acute, outpatient health / mental health / genetic services
General practitioners / primary health networks
Disability service providers
Private providers of health, allied health and wellness services
Non-government organisations
Home care services (e.g. Silver Chain Group – WA, Avivo)
Neurological Council of WA (NCWA)
Housing / accommodation services
Residential aged care facilities

Figure 1. Mapping Huntington's Disease services in WA across the continuum of care & trajectory



Principles of care

The Community of Practice Framework is underpinned by 10 principles of care that were developed in consultation with and endorsed by member agencies of the Community of Practice. As such, they represent consensus opinion of those with specialised knowledge and expertise working in the Huntington's Disease field in WA.

The principles are not intended to be exhaustive or prescriptive. Rather, they have an educative focus, guiding appropriate standards of care and ethical and reflective practice.

The principles strongly endorse the protection, empowerment and support necessary to optimise the health, wellbeing and quality of life of people living with Huntington's Disease. The principles of care and areas of focus are grounded in evidence and supported by local case scenarios. This seeks to define and contextualise each principle in relation to Huntington's Disease.

Principles of care

Holistic and personalised	Huntington's Disease requires a multi-disciplinary approach in order to meet the holistic needs of individuals and their families. Given the complexity and heterogeneity of Huntington's Disease, person-centred and family-centred goal settings are critical to ensure that intervention and support services are tailored to meet an individual's needs as they evolve over time. Care should be concerned with the physical, cognitive, emotional, social and cultural needs of people living with Huntington's Disease. Individuals should be supported to achieve self-identified goals and personal aspirations with access to responsive services and support (e.g. allied health, community support, assistive technology). A service culture and language that makes a person feel valued, important, welcome and safe is essential.
Family centred	Huntington's Disease is a disease of families, acknowledging that every member of the family is affected in some way. Even when family members do not take on a caregiving role, there may still be the need for adjustment. Families should be viewed as whole units, with patterns of relationships that are established over time. It is important to consider the unique and collective support needs of individuals with Huntington's Disease and their family members across the continuum of care. This includes taking time to listen to the stories of families and using this understanding to frame interventions and services (Kazak, 1997). As such, relationships are central to the delivery of care and intervention and support for individuals with Huntington's Disease. Services must be designed and delivered to support and empower strong, confident and capable families. This should support the family to reorganise itself at key transition points along the Huntington's Disease journey.
Lifespan approach	Given the long trajectory of Huntington's Disease it is recognised that there is ample opportunity to plan and choreograph the care and support services required to meet the needs of individuals with the condition and their families (Dawson et al., 2004). A lifespan approach includes planning ahead to ensure appropriate support at critical transition periods, while acknowledging the unique and significant needs of children and young people living at risk of Huntington's Disease. This requires adoption of a palliative care approach and quality of life focus across the continuum of care – aiming to prevent and relieve suffering through early identification, assessment and treatment of pain and other problems (physical, psychosocial and spiritual) that may present.

Wellness and enablement	Living well with Huntington's Disease should be part of the conversation at all stages of the Huntington's Disease journey. Wellness is the process of creating and adapting patterns of behaviour that lead to improved health in the wellness dimensions and heightened life satisfaction (Swarbrick, 2006). This approach aims to generate internal motivation, optimism and a belief in one's own capacity to exert personal control in managing total health needs (Swarbrick, 2006). Individuals with Huntington's Disease should be empowered to learn how to manage problems and stress in order to prevent a crisis situation and maintain wellness. To be successful, the individual's interests, strengths, and potential to achieve personal goals must be understood and central to care and treatment planning. The individual should be supported to establish and maintain a predictable and self-defined daily routine to maintain overall wellness. Where difficulties are encountered, proactive strategies and supports should be established that enable participation and maximal independence, autonomy and quality of life across all stages of Huntington's Disease. This ensures that service delivery is person centred and participation focused; not task or system orientated. A wellness and enablement approach also focuses strongly on early intervention and support, while working to directly address the stigma and discrimination associated with Huntington's Disease through advocacy services.
Supported decision making	Individuals with Huntington's Disease and their families should be actively engaged in care planning, with provision of accessible information and education to support decision making across the continuum of care. This recognises the need for all parties to provide the assistance that an individual with Huntington's Disease requires in order to exercise their legal capacity on an equal basis with others in all aspects of life (Gooding, 2013). Full and accurate assessment of a person's decision-making capacity is required to direct personal care plans and ensure that an individual's ability to input into their own plan is maximised. Furthermore, this seeks to empower individuals with Huntington's Disease and their families to advocate for high standards of care and to combat stigma and discrimination. Care should respect the perspectives and wishes of each individual person living with Huntington's Disease.

Flexibility	Health, care and support services should be flexible to accommodate the unpredictable course of Huntington's Disease and the complexity and uniqueness of each individual's experience (Giles & Lewin, 2008). An individual's needs continuously evolve over time, requiring ongoing assessment and timely access to tailored care related to clinical management of the disorder and its symptoms, community and home support services (e.g. care packages, transport, equipment, housing modifications, assistive devices) and access to suitable respite services (Dawson et al., 2004; Giles & Lewin, 2008).
Predictability	Supporting and caring for individuals with Huntington's Disease requires a careful balance between flexibility and predictability. Given the cognitive changes associated with the disease, individuals with Huntington's Disease can become more rigid, requiring routine, stable and predictable care environments. They can find dealing with change and life transitions extremely difficult, which can be associated with behaviours of concern such as aggression. People with Huntington's Disease cope better when change is gradual, scaffolded and talked about. This requires case coordination, adequate training and continuity of support workers.
Hope and resilience	Living with Huntington's Disease takes energy and strength, highlighting the critical need for practical and emotional support across the continuum of care (Dawson et al., 2004). This highlights the importance of strategies to keep individuals with Huntington's Disease actively involved in their community and engaged in meaningful life roles and activities at all stages of the Huntington's Disease journey. In particular, this requires a focus on strengthening relationships and companionship and providing opportunities for social connection. In some instances, formal psychological support may be required. Support groups, retreats and day centres provide opportunities to foster resilience, connection and coping for individuals, their family and carers (Dawson et al., 2004).
Legacy	Given the progressive nature of Huntington's Disease the principle of legacy should govern care at all stages of the disease. This involves adopting person-centred approaches to care, understanding and celebrating the individual's life history and helping to protect personhood and identity. Life story work should be embedded into care from the point of first contact with the health system. Establishing a strong sense of legacy is expected to support individuals with Huntington's Disease and their families as they move into the more advanced stages of disease, including end-of-life care.

Support for carers	Adequate supports must be provided to assist family caregivers to deal with change, manage competing responsibilities and stressors, find and utilise resources, and provide both physical and emotional care (Williams et al., 2008). Supports for family carers should be in place early, taking a proactive focus, rather than being available only in a crisis situation. This includes providing targeted supports at the recognised transition points along the Huntington's Disease trajectory. To provide appropriate support to family carers, health professionals require an intimate understanding of the unique and complex needs of individuals living with Huntington's Disease and their families. This ensures targeted and responsive tailoring of support groups, weekend retreats, workshops, and respite services to the needs of individuals touched by Huntington's disease. Discussion of carers' mental health should be a routine component of service delivery, along with provision of appropriate and proactive support services.



Key areas of focus

To optimise the health and wellbeing of individuals living with Huntington's Disease and their families in WA, eight key areas of focus have been identified within the Community of Practice Framework. These areas will guide future collaborative and strategic initiatives of the Community of Practice. This represents a shared agenda for change to drive improvement across all systems, services and communities that people with Huntington's Disease come into contact with.

Ensuring people with Huntington's Disease are active and informed agents in their own care journey

Ensuring that people with Huntington's Disease are involved in their care planning and empowered to direct their own journey with Huntington's Disease can help increase compliance with therapy, maintain confidence and sense of control, and optimise participation in society. Doctors, health professionals and service providers should actively engage people with Huntington's Disease and their families in all decisions about their care, seeking to increase understanding of the disease and of factors that can optimise function, quality of life and wellbeing. This includes understanding an individual's unique perspectives and preferences, while acknowledging the changeability of life and the disease journey. Furthermore, service providers must be open to different opinions and preferences, respecting the rights of individuals with Huntington's Disease, particularly in relation to complex ethical issues such as the right to have children and to be a parent. This respects and supports each individual's unique journey with Huntington's Disease. Resources should be available to individuals and their family members to help them know the right questions to ask and the types of supports or services to expect at different stages of the Huntington's Disease journey.

Individuals with Huntington's Disease and their families face an uncertain future, and a future that is often perceived as bleak and hard to confront and control (Dawson et al., 2004). Fears about the future involve dealing with physical and possible mental deterioration, and increasing dependency. Health services must make concerted and committed efforts to facilitate hope, while optimising an individual's sense of control and autonomy.

Factors that have been found to influence quality of life with Huntington's Disease include perceived losses of work, standard of living, social activities, privacy, their home and normal family life (Dawson et al., 2004). Practitioners and service providers should make an effort to understand what gives life quality, meaning and purpose to individuals living with Huntington's Disease, to ensure that tailored intervention and support services can be designed and delivered.

A culture of wellness and enablement represents a philosophy of care focused on helping individuals to achieve their maximum potential with reference to their own health and social wellbeing (Dewing, 2004). This is a shift from care that focuses heavily on risk assessment, prevention of risk and a culture that makes people more dependent

in multiple ways and results in 'de-skilling' or excess disability (Dewing, 2004).

Health professionals play a critical role in facilitating independence and autonomy, moving from 'doing to', to 'doing with' people with Huntington's Disease – enabling them to do as much as possible for themselves. This requires direct conversations and negotiations with individuals living with Huntington's Disease and their families based on their values, preferences and life story. Knowing the person and being in an enabling relationship is skilled work, demanding expertise (Dewing, 2004). Health professionals, support workers and family carers must be educated, trained and supported to provide enablement and reablement services. Such services must be delivered in the context of a multi-disciplinary team, driven by goals tailored for the individual and focused on functional participation, wellbeing and quality of life outcomes.

Documenting individual preferences, goals and care plans across the continuum of care

To optimise holistic and consumer-directed health care, and integration and coordination of care across agencies, clear and effective communication between service providers is important. This should include the use of personal wellness and care plans that follow the individual with Huntington's Disease across the continuum of care. This could function as a health journal, while facilitating important conversations between the person with Huntington's Disease and their health care providers. It is important that care plans evolve with the individual's changing circumstances and preferences over time.

Planning for the future and starting difficult conversations early

The long duration of Huntington's Disease and understanding of the natural history of the disease provide a window of opportunity for forward planning, supporting individuals with Huntington's Disease to become active agents in their own care and plan for the future. Providing psychological support and accessible information about Huntington's Disease and available care and support options to the individual and their families is essential to coping with life following diagnosis and the progression of Huntington's Disease over time (Dawson et al., 2004).

Planning for the future may help reduce the impact of transition periods or unavoidable negative events, serving a protective function. This may help to prevent an individual starting on a pathway with negative outcomes. Planning for the future also helps to ensure that future financial needs can be met (Dawson et al., 2004). By starting difficult conversations early, more opportunities to understand and document individual preferences and perspectives are available, promoting person centred and proactive care.

It is noted in the Huntington's Disease literature that families can be reluctant to plan for the future and talk about 'what ifs' (Dawson et al., 2004). However, if these

conversations do not take place, fragile family and support systems can crumble in the face of health threats (Zarit, 2001). This supports a case management approach, where readiness for information can be monitored. The amount, timing and type of information should be carefully and sensitively considered (Dawson et al., 2004). It is recommended that information is tailored and filtered for individual families (Dawson et al., 2004) and re-visited on an ongoing basis.

Conversations should explore preferences and decisions about medical treatment a person wishes to receive in the future, including a systematic approach to advance care planning. This can help to minimise or prevent significant distress for families faced with difficult ethical decisions in the advanced and end stages of the condition. This should encourage open conversations about end-of-life issues and foster positive attitudes about death and dying (Kristjanson, Toye, & Dawson, 2003).

Embedding a palliative care approach across the continuum of care is required, aiming to improve quality of life and resilience (Kristjanson et al., 2003). This encourages a focus on pain and symptom management at all stages of Huntington's Disease, while ensuring that the physical, psychological, spiritual and social challenges facing patients and their families are considered and supported (Kristjanson et al., 2003). Conversations should also consider future housing and financial needs so that plans can be initiated in advance.

Working with families and supporting family carers

A diagnosis or family history of Huntington's Disease is known to have significant repercussions for the whole family due to the hereditary nature of the condition (Dawson et al., 2004; Vamos et al., 2007). Siblings experience survivor guilt if they receive a negative test result when other family members have received a positive result (Dawson et al., 2004). Fear for the future and for the next generation is common (Dawson et al., 2004) and it is also possible that one family member may affect one or many others with his or her own emotional issues (Vamos et al., 2007).

Huntington's Disease can have a devastating effect on marital and wider family relationships, and young people have described how Huntington's Disease 'broke their family apart' (Keenan et al., 2009). It has been noted that terminal illness like Huntington's Disease can exaggerate existing family structures and coping styles, with "cohesive families becoming more cohesive and disorganised ones more chaotic" (Vamos et al., 2007, p. 400). This highlights the critical need to approach Huntington's Disease from a family perspective when assessing health and psychosocial support needs.

Family caregivers of people with Huntington's Disease have reported multiple aspects of emotional distress (Williams et al., 2008) and can experience exhaustion, isolation and burden associated with the responsibilities of care (Dawson et al., 2004). This can compromise the wellbeing of family carers (Williams et al., 2008) and highlights the need for tailored education, counselling and ongoing psychological support as a fundamental standard of care (Dawson et al., 2004). This stress for family carers can

be compounded in the context of Huntington's Disease, often involving the additional stress of other family members undergoing genetic testing and facing a diagnosis (Dawson et al., 2004). Adequately supporting family carers can reduce the risk of premature institutionalization and help families to plan for the future. This should focus on strengthening resilience and hope within the family system.

It is important to note that not all individuals with Huntington's Disease will want family members involved directly in their care and management. This should be considered and respected.

Supporting children and youth

Children and young people living within Huntington's Disease families have unique and significant needs that service providers must respond to. Youth report experiencing feelings of helplessness, anger and isolation that can be associated with acting out behaviour, self-harm, and use of alcohol and drugs (Keenan et al., 2009). Huntington's Disease is associated with stigma and taboo, which can make family conversations about Huntington's Disease challenging.

Family rifts, secrecy and poor communication are common. Historically, youth services have not been available for young people living in Huntington's Disease families; however, the need for tailored youth services is being increasingly recognised. This acknowledges that early childhood experiences and protective relationships have long-lasting effects on brain and psychosocial development. Timely and effective identification and management of risk factors is critical to minimise the negative impacts on children's and young people's safety, stability and development; while acknowledging the complex interaction between risk factors and protective factors (Department of Disability, Housing and Community Services, 2012).

Research has not adequately explored the experiences of children and young people finding out about a genetic illness or Huntington's Disease being in their family or what this means for them (Keenan et al., 2009). Currently, there is no consensus opinion on the optimal age for these conversations to take place (e.g. early, mid or late adolescence) (Holt, 2007; Van der Meer et al., 2007). There is evidence to suggest that young people sense 'something is wrong' long before they are told, even when as young as five (Easton, 2003) and do seek explanations. Research also suggests that children can cope with knowing, even at a young age (Keenan et al., 2009) and that children who suspect that something is being kept from them can experience feelings of anxiety and confusion (Keenan et al., 2009). Triggers for 'needing to tell' include a relative's death, parental divorce, career/exam choices, and first relationships; or the time when telling begins to outweigh the benefits of not telling (Keenan et al., 2009).

Parents living with Huntington's Disease should be supported to make such decisions around disclosure to their children with access to tailored information and counselling services. It appears that disclosure should not be a one-off event, but rather a process that unfolds incrementally over time and that should be negotiated at an individual child's pace.

Care needs to be taken in the manner and style that information is delivered to youth. When disclosure is made this should be an educative process. This requires the support and guidance of practitioners and providers with an intimate understanding of Huntington's Disease and its impact on children, who can support both parents and young people through this complex process. Individual preferences relating to the process and timing of disclosure and patterns of communication should also be respected and supported by practitioners and providers. The significant 'disclosure burden' that parents carry should also be acknowledged, representing another source of anxiety or worry for people living with Huntington's Disease and their families.

In some instances, limited parental disclosure acts as a protective factor for children; however in other instances it can increase the experience of isolation and helplessness. This requires close monitoring of the child at the time of disclosure and access to tailored support services for young people at risk of or touched by Huntington's Disease. Research indicates that young people go through stages of coping and not coping, with different levels of engagement with their own personal risk (Keenan et al., 2009; McAllister, 2002). Research also indicates that children can be active agents in their own learning about Huntington's Disease, actively seeking out or listening for information. Different modes of support for young people should be available, including use of social media.

Support services should aim to identify and bolster protective factors, for example, social skills, attachment to family, independence, social networks and participation in positive and supportive community environments (Department of Disability, Housing and Community Services, 2012). The critical need for dedicated youth services for children growing up in Huntington's Disease families cannot be overstated, aligning with family centred and lifespan approaches to the integrated management and care of Huntington's Disease.

Supporting transition and critical periods along the Huntington's Disease trajectory

A number of transition or critical periods have been identified along the Huntington's Disease timeline or trajectory. This includes the time when risk of Huntington's Disease is disclosed or realised and when decisions around whether or not to proceed with predictive testing are being made. Critical periods may align with key life stages, such as deciding whether to start a family of one's own or when making decisions about career options and relationships. Another transition period occurs at the time symptoms start to emerge and diagnostic testing is required. Further critical periods may be associated with the progression of symptoms, where the functional consequences of Huntington's Disease become more prominent, where activities become more restricted (e.g. driving, managing finances, capacity to work) or the level of dependence on others starts to increase (Paulsen et al., 2005). A necessary move into residential aged care and progression into the advanced or end stages of disease represent other transition periods.

It is important that practitioners and service providers understand and recognise these critical points along the Huntington's Disease timeline and feel equipped to provide adequate information and support to minimise negative impact. This links to 'starting difficult conversations early' and planning for the future to prepare for transition periods and ensure that additional, tailored supports are available. At each transition point, individual preferences, wishes and decisions must be respected.

Transitioning into the advanced stages of disease, there is a risk that individuals with Huntington's Disease may be over-nurtured, with restricted opportunities to participate and take negotiated risks. The need to support the person with Huntington's Disease to 'live the good life' into the end stages of the disease should be considered by service providers.

Ensuring highly skilled and capable health professionals and care workers

It is of paramount importance that health professionals and service providers demonstrate expert knowledge about Huntington's Disease and the recommended principles and standards of care (Dawson et al., 2004). When individuals with Huntington's Disease and their carers have to access the hospital system and mainstream health services they report that health professionals have a lack of knowledge about Huntington's Disease (Dawson et al., 2004). This is associated with a lack of trust in the standard of care provided, which is detrimental to health and wellbeing outcomes. Similarly, to ensure satisfactory respite services it is of paramount importance that health professionals are well educated and that services can be trusted to provide excellent standards of care (Dawson et al., 2004).

Organising residential respite care can be particularly problematic when there are behavioural or management issues (Dewing, 2004). There is a need for respite and long-term care to be provided in locations where all staff have training and skills to deal with the complex care needs of people with Huntington's Disease (Dawson et al., 2004). Particular expertise is required in the areas of behaviour management, family support, nutrition, swallowing, and communication, as well as Huntington's Disease friendly environments (Dawson et al., 2004).

The Community of Practice Framework and Principles of Care should be used to design education and training material, ensuring that individuals, teams, and organisations possess the underlying core values, knowledge, attitudes, skills, and capabilities to support individuals with Huntington's Disease and their families. This includes capabilities to promote autonomy and consumer-directed care, ability to identify and focus on strengths and individual potential, and a commitment to holistic, person- and family-centred philosophies of care. Furthermore, practitioners and service providers should know how to help people with Huntington's Disease and their families find the right combination of services, treatments and supports that they require at any point in time, eliminating barriers to participation in meaningful life roles and activities and optimal wellbeing.

Fostering Huntington's Disease-friendly environments

Huntington's Disease friendly environments must provide a calm predictable environment and establish a routine for the day that an individual is comfortable with. It is important to understand any trigger factors that may influence a person's mood and behaviour. Environmental supports, activity and information should be tailored to an individual's level of function and capacity. The environment should not overload the person with information or stimulation. The complexity of the environment should be minimised, keeping things in obvious and consistent places. Tasks should be broken down into smaller, achievable steps, enabling the individual's performance and maximising independence and participation. Effective communication and supported conversation strategies should be embedded into routine care and interactions. This should provide clear, simple instructions and time for the person with Huntington's Disease to process information and respond. External memory aids such as clocks, calendars, whiteboards and activity schedules may assist memory and executive difficulties.

Environmental risks should be assessed on an ongoing basis. Risks may include physical / falls risk, personal risks related to smoking and other decisions, risks associated with nutritional status and swallowing, and risks associated with community access and use of public transport. When managing risk the perspectives and wishes of the person with Huntington's Disease are paramount, requiring an understanding of and commitment to 'negotiated risk'. This relates to the rigid thinking, reduced flexibility and difficulty dealing with change that may be experienced by individuals with Huntington's Disease.

Huntington's Disease friendly environments should enable individuals with Huntington's Disease and provide continued opportunities for learning and continued personal growth at all stages of the Huntington's Disease journey.



Outcomes of the Community of Practice Framework

The next step is to embed the framework and principles of care into everyday practice, ensuring positive outcomes for individuals, families, and systems. The Community of Practice Framework guides clinicians to monitor and improve the health and wellbeing of people with Huntington's Disease across the continuum of care and at critical transition periods in the Huntington's Disease journey. The framework is aspirational, however, it also provides specific direction for routine clinical care and service provision. This will inform selection and development of assessment and outcome measurement tools to identify and monitor needs over time. The framework emphasises the need for wellbeing and quality of life to be measured across all stages of the Huntington's Disease trajectory.

Outcomes at a broader system level should provide evidence that services are responsive to the needs of individuals living with Huntington's Disease and their families, choices for treatment and care are available, services are delivered in a timely and proactive manner, the rights of people with Huntington's Disease are respected, protected, and promoted, and enabling services and communities are delivered and achieved. It is hoped that the framework will foster new and innovative service designs and care initiatives.

Implementing the Community of Practice Framework

The Community of Practice Framework has been endorsed by core agencies delivering care to individuals with Huntington's Disease in WA. It will be used to guide consultation and liaison across agencies and to set strategic priorities. This aims to support continuity of care so that individuals with Huntington's Disease can move between service elements as their needs change and receive appropriate, high quality care at any time.

Furthermore, the framework will be used as a mechanism to plan, evaluate and improve services as part of a continuous service development and quality assurance process. Service providers are also encouraged to use the framework to develop local protocols for care. Huntington's Disease champions should be identified across agencies to act as opinion leaders and mentors to ensure that the principles of care are upheld.

The framework will be regularly reviewed and updated in light of new evidence and policy and practice changes into the future. Training packages based on the principles of care should be developed. Furthermore, clinical resources (e.g. assessments, goal setting and care plan templates) should be identified and developed that further support implementation of the Huntington's Disease Community of Practice Framework. In the future, standards of care or recommended care practices could be developed based on the principles of care and best available evidence. Furthermore, key performance indicators and performance standards should be articulated that align with the principles of care, together with monitoring and evaluation mechanisms.

The dissemination and sharing of successful service initiatives applying the framework and principles of care are strongly encouraged at a local, national and international level.

Case study

Joan is a 42-year-old woman with symptomatic Huntington's Disease living with her 17-year-old daughter in the community. She was referred to the Neurosciences Unit Huntington's Disease clinic in 2002 by a Neurologist (Royal Perth Hospital) for psychiatry, speech pathology and social work services.

Joan had previously worked as a cleaner and itinerant work as a shearer's chef. She had a history of alcohol and drug abuse and periodic verbal abuse and threatening behaviour towards others, including health professionals. She had subsequent periods of homelessness and itinerancy, living in her car. She had intermittent accommodation with a former school friend, punctuated by domestic violence. Her car was later written off and she then spent periods of time sleeping at the beach or in toilet blocks. She refused crisis accommodation and women's refuges as she did not want to be separated from her dog. She was increasingly coming to the attention of members of the public, local MP, park rangers and the police.

Joan became increasingly estranged from her daughter over time due to her presentation. She had one brother symptomatic with Huntington's disease and another brother who attempted to support her.

Joan's level of insight into her accommodation and care needs declined over time. While she acknowledged her Huntington's Disease, she neglected her health and dental care and was increasingly at risk to herself and others, due to volatile behaviour. The level of financial and welfare assistance and crisis intervention due to her circumstances increased over time, alongside progression of her Huntington's Disease and decline in cognitive and physical ability. She presented with increased paranoia, falls risk and choking episodes.
An involuntary admission to Graylands Hospital was recommended due to risk to herself and others.

Specific scenario

Joan was living in a toilet block on a Perth beach with her dog. She was found by the ranger soaked by the morning sprinklers with an empty bottle of spirits beside her. Adult mental health service declined to progress an involuntary admission to hospital, advising that it was a lifestyle choice. She was subsequently admitted to Graylands hospital on the third attempt to successfully organise an involuntary hospital admission. Joan's goal was to live independently in the community with her dog and to die at home. She had significant fear of residential aged care, due to witnessing her mother and sister dying from Huntington's Disease in care. Joan was fiercely independent and this needed to be respected in all contact with her – seeking ways to optimise her autonomy and sense of control.

This scenario illustrates the complexity of issues associated with Huntington's disease. Achieving a positive, safe and acceptable solution required skilled case coordination and management that strongly upheld principles of the Community of Practice Framework as outlined in the Table below.



Holistic and personalised	The management plan needed to be highly responsive to Joan's situation, her personal aspirations and psychosocial needs in order to maintain an effective therapeutic relationship with her. This adopted a person-centred approach within the parameters of Joan's assessed cognitive capacity. Personalised care required intimate understanding of the nature of Huntington's Disease in order to respond to and advocate for her needs, while respecting her fierce independence and previous lifestyle – that had always been itinerant in nature. Coordinated and targeted efforts were made to show Joan that her care was being tailored to meet her unique and specific needs. For example, Joan was thrilled to have her "own chef" as a result of a speech pathologist from the Neurosciences Unit providing dysphagia education to care staff and appointing a key worker to coordinate meals.
Wellness and reablement	A wellness and reablement approach was central to care and support provided to Joan, assisting her to adopt healthy lifestyle choices. This was incredibly challenging in light of her alcohol and drug abuse, history of domestic violence and self-neglect (e.g. dental care). A key strategy for enhancing wellness was linking Joan to the Huntington's WA Day Centre along with social support from a specialist Huntington's Disease Advisor. These strategies had a profound impact on Joan. She thrived on the social connection provided by the highly targeted Day Centre activities and home visits.
Hope and resilience	Engendering a sense of hope for the future in terms of attempting to secure acceptable accommodation and care to meet Joan's goals was essential, while balancing this with realism and avoiding false assurances that this would eventuate. Adequate safeguards were required given that Joan would have been at serious risk of harm living in the community without adequate supports and funding given her advanced Huntington's Disease. Her case manager sought funding from the Department of Communities (Disability Services) to provide a full package of care to the client in her own home to include community access and provision of information and education on Huntington's Disease to the service provider. Home modifications were arranged to optimise safety.

Predictability	The client was supported to live independently in the community despite her advanced Huntington's Disease and cognitive impairment as a result of the package of care and social supports in place. In this highly supported environment positive daily routines could be established that were responsive to Joan's needs and respectful of her wishes
Support for carers	Support for Joan's formal and informal carers was an essential part of her management. Psychoeducation was provided to care providers and her brother (next of kin) on the presentation and management of Huntington's Disease. This focused on helping them to understand the challenging and responsive behaviours that Joan presented with. Care workers were also rotated regularly. Joan often "sacked them" but reinstated them later.

Key learnings from this case scenario

Overall, the case strongly illustrates the importance of engaging individuals with Huntington's Disease as active and informed agents in their care, to support transitions and critical periods along the Huntington's Disease trajectory, and ensure that health professionals and care workers are highly skilled and equipped to manage the complexity of issues that can arise.

The need for self-determination shines through this case study, despite the significant obstacles for clinicians and service providers in achieving this. It also highlights the importance of the lifespan approach to Huntington's Disease, addressing long-term issues and evolving care as needs change over time. This requires establishing long-term therapeutic relationships that can be volatile given the nature of Huntington's Disease, respecting the impact of cognitive and psychological symptoms of the condition.

Services should be designed around such challenges, rather than seeing them as barriers to care (e.g. rotation of care workers, assignment of key workers with a strong relationship to the individual client). This requires flexible provision of services and strong collaborative practice built upon intimate understanding of the complex nature of Huntington's Disease and how best to manage challenging behaviours. Through enacting the principles of the community of practice behaviour, the challenging behaviours that Joan presented with were successfully managed allowing a positive outcome to be achieved.

Outcome

Joan died peacefully at home in 2014 in a setting that was reflective of her personal wishes and preferences. This demonstrates that despite the enormous complexity of Joan's journey with Huntington's Disease, it is possible to achieve highly positive outcomes for the individual, their family and care systems.

Glossary

Early intervention:
Aims to prevent excess disability or unnecessary decline and avoid or delay the need for higher level of care.

Enabling approach:
Is "doing with" rather than "doing for" the individual with Huntington's Disease, in order to enhance autonomy, independence and participation.

Family systems approach:
Acknowledges that a change in one family member will influence the entire family system. This views the family as a complex, dynamic and interactive unit. An individual with Huntington's Disease cannot be understood without a careful examination of the parts of his family and relationships.

Lifespan approach:
Focuses on the holistic (i.e. physical, mental, social, emotional and spiritual) needs of the individual living with Huntington's Disease and their family from childhood through to the end stages of the condition – responding proactively to evolving needs over time.

Palliative care approach:
A palliative care approach is distinct from specialist palliative care services. A palliative care approach has a broader focus, aiming to improve the quality of life of individuals facing life-threatening illness and of their families, by preventing and relieving suffering through early identification, assessment and treatment of pain and other problems (physical, psychosocial and spiritual). A palliative approach encourages more open communication about end-of-life issues.

Person-centred approach:
Places the individual with Huntington's Disease at the centre of all decision-making about their life (aspirations, goals) and the supports and/or services that they require.

Restorative / reablement:
Restorative or reablement services represent short-term, multi-disciplinary interventions for individuals with Huntington's Disease at risk of unnecessary or preventable decline. When successfully targeted and implemented such services can delay, reduce, or remove the need for longer-term or ongoing support.

Strengths-based approach:
Focuses on what the individual with Huntington's Disease can do and what they want to be able to do; rather than focusing on loss or weaknesses.

Wellness approach:
Encourages and enables individuals with Huntington's Disease to make healthy and more satisfying life choices (e.g. exercise, diet, nutrition, cognitive stimulation, social participation) that optimise health and wellbeing.

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Contributing organisations

Huntington's WA

Neurosciences Unit (NSU), North Metropolitan Health Service, Mental Health

Brightwater Care Group

MSWA

Department of Communities (Disability Services)

South West Huntington's Network

