Submission in Relation to the Accommodation needs of Younger People with Neurodegenerative Disorders – Huntington Disease

Hunter Genetics is a unit of the Hunter New England Health Service providing a general genetics unit and specialty areas such as the Fragile X, Hunter Family Cancer Service and The Huntington Disease Service. Approximately, 1000 new families are seen each year, with approximately 700 from the local Hunter area, the remainder from outreach clinics conducted at Central Coast, Taree, Port Macquarie, Coffs Harbour, Lismore/Tweed/Grafton, New England, Dubbo/Mudgee, Muswellbrook and Broken Hill.

The Huntington Disease Service

Clients are referred to the service for specialised treatment and/or management. The Huntington Disease Social Worker receives referrals from general practitioners, neurologists, clinical geneticists, genetic counsellors, and other social workers for counselling and management of the psychosocial issues influencing individuals and families affected by Huntington disease (HD). There are a range of complex human problems that accompany the diagnosis, onset, and course of an inherited neurological disease like HD. The Huntington Disease Social Worker also co-ordinates the presymptomatic gene testing program for HD and arranges the clinic for the liaison psychiatrist who provides psychiatric support at a monthly clinic to manage emotional and behavioural sequelae associated with Huntington disease.

Huntington Disease

Huntington Disease is a progressive neuropsychiatric genetic disorder. It is inherited in an autosomal dominant fashion, which means that it occurs equally in both males and females, and can be inherited from an affected mother or father. The faulty gene causes irreversible and untreatable progressive damage to brain cells in the basal ganglia frontal lobe and elsewhere.[1]

Huntington Disease usually appears between the ages of 35 and 40 years. *The major symptoms include movement disorder, personality disorder and mental deterioration*. Huntington Disease tends to run its course from diagnosis to death over an average of 15 - 20 years.

Characteristically there is an organic dementia with progressive impairment of memory, loss of intellectual capacity, apathy and inattention to personal hygiene. Early in the disease there are usually irritability, impulsive behaviour, bouts of depression or episodes of violence. In some patients there is also frank psychosis – usually requiring admission to a psychiatric centre and/or outpatient psychiatric management with psychotropic medication.

Commonly, the person with Huntington Disease will experience the following changes;

- Eating and Swallowing - Difficulties with eating and swallowing (dysphagia) and maintaining constant body weight are among the most troublesome

[1] Summary of Symptoms of Huntington's Disease
From DR J Snowden – Taken from HD Newsletter of England and Wales. Issue no.5 Winter 1996.

complications of Huntington Disease. Factors which lead to these problems, include: changes in appetite; choreiform movements – problems of chewing and facial control while food is in the mouth; deterioration of the muscles involved in swallowing, changes in mood, difficulty eating in the presence of involuntary movements of muscles in the throat, larynx and chest wall involved in breathing. Eventually this progresses from dysphagia to aspiration of food and pneumonia because of involuntary inward breaths while food is in the airway.

- Speech and Language Difficulties – speech production disturbances begin early on in the disease process. As the disease progresses, speech often becomes unintelligible and individuals become non-verbal in the advanced stages of the disease. Clear communication eventually becomes almost impossible even though the frustrated patient knows what they want to say.
- Chorea is the major motor sign of Huntington Disease and can be defined as "A state of excessive, spontaneous movements, irregularly timed, randomly distributed and abrupt. Severity may vary from restlessness with mild, intermittent exaggeration of gesture and expression, fidgeting movements of the hands, unstable dance-like gait, to a continuous flow of disabling, violent movements. Choreic movements are continuously present during waking hours, cannot be voluntarily suppressed by the patient, and worsen during stress." These continual movements are one of the reasons people with Huntington Disease require a carbohydrate loaded diet and specific weight maintenance dietary supplements.
- Frustration, irritability and loss of self-confidence may occur to some extent as an understandable reaction to the presence of a disabling illness and to the loss of communication and personal independence the disease entails.
- Loss of Drive and Initiative, Huntington Disease affects the subcortex of the brain, the area responsible for drive and initiative. This part of the brain allows us to think, plan and generate actions to solve problems. It also provides the persistence to complete a task. When this part of the brain is damaged, the drive, or foresight to independently initiate activity is severely compromised, even though in the early stage the skill to carry out the activity is retained. It is not uncommon for Huntington Disease sufferers to carry out everyday tasks less efficiently than before. The sufferers lose the capacity to self-monitor efficiency or efficacy and needs a supervisor to remind them of the next step in multi-step tasks like dressing, bathing, shaving, cooking or eating.
- Mental Flexibility – sufferers can seem inflexible and mentally rigid. They may adhere to set behaviour patterns or routines and appear unwilling to adapt to new situations, or altered circumstances. They generally feel most comfortable or confident in highly familiar situations, involving a fixed routine
- Hygiene & Self-Care - It is very common for a decrease in interest in personal appearance and hygiene to occur. Huntington Disease impairs personal and social awareness and blunts emotions. Personal insight decreases and thus the sufferer does not feel ashamed. It is important to gently motivate appropriate personal hygiene, once this is necessary it requires daily supervision
- Depression – is relatively common, although the loss of drive and initiative does not necessarily indicate a depressed mood. Nevertheless, depression should always be considered, particularly if the change in the patient's level of motivation and interest has occurred relatively rapidly
- Irritability and Aggression – although some Huntington Disease sufferers may be relatively even tempered and somere even euphoric, it is not uncommon for sufferers to become emotionally volatile. They may flare up

over trivial issues. Patients often feel a sense of internal agitation and are aware that they are easily annoyed; yet sudden outbursts of anger come without warning and are generally outside their control.

- Adjustment / Denial – the onset of Huntington Disease leads to major changes to lifestyle for both the sufferer and their family. This can be made more difficult if the sufferer refuses to accept that there is anything wrong. It is easy to assume the changes are as obvious to the patient as to other people. However, research shows that Huntington Disease sufferers may not have normal experience of their involuntary movement. *In addition, the condition can affect the ability to self-monitor; thus insight into the problem is impaired.* Refusal to accept illness is not simply obstinacy on the part of the patient; it is a feature, which occurs in some (not all) sufferers as a consequence of the disease process itself. Loss of insight can lead to disastrous personal, financial and professional decisions.

The behavioural changes noted above are an integral part of the condition, arising as a direct consequence of physical changes, which take place in the brain.

Shoulson [2] defines five stages of Huntington Disease and it usually in the latter two stages that patients with Huntington Disease are no longer able to live safely in the community even with the support of community based services, access to which is steadily becoming more restricted. Maintaining ongoing independence is the goal of all Huntington Disease services, however, due to the degenerative nature of the condition, the devastating nature of the symptoms of Huntington Disease and the lack of a cure in the foreseeable future, most will end their life in a residential (aged care) facility.

Stage	**Engagement in Occupation**	**Capacity to handle financial affairs**	**Capacity to handle domestic responsibilities**	**Capacity to perform activities of daily living**	**Residence**
Stage 1	Usual level	Full	Full	Full	Home
Stage 2	Lower level	Requires slight assistance	Full	Full	Home
Stage 3	Marginal level	Requires major assistance	Impaired	Mildly impaired	Home
Stage 4	Unable	Unable	Unable	Moderately impaired	Home or extended care facility
Stage 5	Unable	Unable	Unable	Severely impaired	Total Care facility only

There are approximately one hundred and thirty people living in the Hunter region alone who are gene positive for Huntington Disease. Fifty-six of these 130 are currently over the age of 30 and are or will soon be showing obvious clinical signs of Huntington Disease.

Currently two are in acute health care facilities waiting long-term placement. Both are assessed as low care by the Aged Care Assessment Team.

[2] Shoulson I. (1981) Huntington's Disease: functional capacities in patients treated with neuroleptic and antidepressant drugs. Neurology 31, 1333-1335. Shoulson and Fahn S. (1979). Huntington's Disease: clinical care and evaluation. Neurology 29: 1-3.

There are approximately twenty-two known clients who will require residential care for their own safety and well being in the next two - three years. Five of these twenty-two are in need of imminent or urgent placement. Not all clients who are gene positive with Huntington Disease are clients of the service or regular recipients of assistance through the Huntington Disease Social Work Service at Hunter Genetics. As a consequence, clients are sometimes placed into residential care directly from hospital, usually as they are assessed as unsafe to continue living in the community.

There is a serious misconception, both in the community at large, and within the health and caring professions that there is purpose built facilities for people with Huntington Disease. The first of these is a four-bedroom house in Mayfield known as "Camellia Cottage," which was officially opened in 1999. The house was funded under the Building Better Cities – Communities and Special needs Housing Project and is required to be compliant with the program's guidelines. Under the original project agreement, the current head lease does not permit the provision of a live in 'manager / caretaker.'

The NSW Land and Housing Corporation own this property and is head leased to the Australian Huntington Disease Association at a peppercorn rental for 10 years until October 2009. The aim of the house is to provide 'transitional' housing for people who are mildly symptomatic for Huntington Disease. The term "transitional" in this context is not the same as "transitional units" that provide care for elderly people awaiting placement in an aged care facility after their discharge from an acute care facility.

Camellia Cottage is an option for people who are currently living alone, or whose family are no longer able to care for them, most commonly due to the breakdown in family relationships. It is not an alternative to institutional living and is not staffed at any time by either a caretaker, or residential care workers. Its primary aim is to provide accommodation in which each individual tenant has services brokered in to meet their individual needs. The house has on occasion provided emergency housing for clients with Huntington Disease who would other wise have been homeless. This is conditional upon the availability of a room and the requirement that the safety and well-being of the other tenants must not be placed at risk. Camellia Cottage is neither a group home, nor is it a boarding house.

Another strongly held misconception is that a purpose built hostel (similar to the unit at Lottie Stewart Hospital in Sydney), exists at Wescott Centre at Stockton (Presbyterian Aged Care). Wescott does have young people as residents, some of whom have Huntington Disease. However, Wescott is an aged care facility and they are not funded to take any more young people. Despite regularly publishing this fact, the misconception remains that *all* young (and older) people with Huntington Disease in the Hunter who require residential care will have an automatic placement at Wescott.

In recent months, two particular cases have caused great ethical concern to the workers involved in the provision of services to these clients.

Case number one "Anna" is a 42 year old woman with advanced Huntington Disease (stage five), who was living in the community until December 2004. In this case it only took approximately 2-3 months to have the approval for a high care ACCR, however, the brain damage caused by the Huntington Disease caused Anna to refuse to consider living any where but in her own home. This was despite grave concerns for her health and wellbeing. The issues that concerned her care workers included;

- Suspected financial and physical abuse
- Anna was often out in the street approaching strangers to assist her to get money from her account
- Anna often sat out the front of her home inappropriately attired (no underwear, wearing a short skirt)
- Frequent falls – observed to fall at least 3 times / hour. Falls resulted in a injuries of varying severity
- Anna frequently forgot to turn the gas off and then smoked in the house, threatening the safety of other tenants in the block
- Anna left windows and doors unlocked leaving her vulnerable to break ins and possible assaults
- Anna was in arrears on some of her utilities
- She refused to consider alternative housing options
- There was a lack of meaningful social activity
- Anna's speech was unintelligible
- She had frequent choking episodes each day
- Severe weight loss – went from ladies size 10 to children's size 6 in 4 months
- Violent outbursts towards children – grandchildren present at same time. Police were called on a number of occasions to intervene.
- Threats of self harm

In an effort to find suitable accommodation for Anna 16 facilities (aged care) were sent standard application forms, with a cover letter requesting she be placed on their waiting list, and a copy of the ACCR. Four facilities declined placing her on the waiting list, as they felt unable to manage her care needs, particularly her behavioural problems. She was not placed until her Guardian for the Guardianship Board placed pressure upon a facility to take her. The entire process of placing Anna took over 3 years.

Case 2: Maria, a 45 year old woman who also is living in the community with the maximum allowable / available community support services is unable to get an ACCR because of:

- "Her age
- She is not dependent enough for high care and would find it difficult to fit in with a low care facility unless it was dementia-specific.
- She needs another type of supported accommodation i.e. small group house with adequate support."

She is ineligible for assistance via DADHC (ACP) as she is unable to manage to arrange the attendants and she has increasing needs over time. In addition, there are no ACPs available in the Hunter at present.

Maria is at stage 4 / 5 of the disease and faces the following concerns

- Up to 8 falls per day – most result in head and face trauma. Mobility problems sufficient that staff require she move around by using a wheelchair for her own safety and that of the staff who visit her.
- Speech and swallowing difficulties – she now requires feeding 3 times per day as she is no longer able to feed herself due to the severity of her choreic movements, and the protrusion of her tongue (another of the involuntary movements).
- Decreased insight – refuses to consider any other facility than Wescott, despite having been advised on multiple occasions they are unable to provide a bed for her.

- Guardianship application in process
- Weight loss from woman's size 14/16 to size 8 in last 4 months
- Unable to remember her own address
- Behavioural problems such as episodes of aggression, refusal to accept medical attention after falls
- No longer capable of using electronic banking (automatic teller), and is no longer able to sign her own name.

The behavioural aspects of Huntington Disease and the need for high level care in the end stage make people with Huntington Disease extremely difficult to place. Although many are still ambulant their balance is poor, leading to collisions with furniture and their frail neighbours in institutions. Aged care facilities need to consider the needs of their other residents and their capacity to manage the often enormously demanding requirements of someone with Huntington Disease, whose extremely poor impulse control makes them prone to outbursts of frustration. The overt choreic movements of Huntington Disease can be tremendously confronting to many people and is often cited as one of the reasons a facility will be unable to take a placement.

For those who suffer the muscle rigidity in the end stages, facilities have to deal with problems of pressure areas. Those that require PEG feeding, of course, also require specialist nursing care to avoid infection. Due to decreasing mobility, many people with Huntington Disease will require specialist seating which costs on average between $2,500 and $3,000. These costs are not factored into the running costs of facilities and make people with Huntington Disease a particularly unattractive candidate for placement in any facility.

The moral and ethical dilemmas faced by health care workers functioning in this area on a daily basis are primarily attributable to non existent purpose-built residential facilities for young people (under 60 years of age) who require more care than can be provided in the community. While it is universally recognised that it is inappropriate to place young people in an aged care facility, the ethical dilemma of placing a young person in an aged care facility is possibly the lesser of two evils when no other special facilities exist. The decision to institutionalise a young person is not lightly considered, and the longer the ethics of placement are debated the greater the danger for the young people involved. Using age as a reason to block provision of accommodation (when none other exists) is obstructive, a breach of duty of care, and in some cases a contravention of human rights.

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