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Submission to Joint Select Committee on End of Life Choices
From The Motor Neurone Disease Association of Western Australia Inc. (MNDAWA)

The Motor Neurone Disease Association of Western Australia Inc. (MNDAWA) respectfully submits this document for consideration in the inquiry into the need for laws in Western Australia to allow citizens to make informed decisions regarding their own end of life choices. In this submission, MNDAWA will specifically address:

- the practices currently being utilised within the health community to assist a person living with Motor Neurone Disease (MND) to exercise their preferences for the way they want to manage their end of life, including the role of palliative care; and
- provide commentary on the role of advanced health directives, enduring power of attorney and enduring power of guardianship laws and the implications for individuals living with MND that may be covered by these instruments in any proposed legislation.

What is Motor Neurone Disease?

Motor Neurone Disease (MND) is the name given to the group of diseases in which the motor neurones undergo degeneration and die. Amyotrophic Lateral Sclerosis (ALS), Progressive Muscular Atrophy (PMA), Progressive Bulbar Palsy (PBP) and Primary Lateral Sclerosis (PLS) are all subtypes of motor neurone disease.

Although MND is the widely used generic term in the United Kingdom, Australia and parts of Europe, ALS is used more generically in the United States, Canada and South America. MND is also known as Lou Gehrig's disease in the US after a famous baseball player who died of the disease. The following key information on the disease is from the peak body website, MND Australia:

- There is no known cure and no effective treatment for MND.
- Each day in Australia two people die from MND. This statistic is based on cause of death certification as MND being the underlying cause. This is separate from those people who have MND, but who die from something else (such as a car accident, cancer or cardiac disease).
- Each day in Australia two people are diagnosed with MND.
- People with MND progressively lose the use of their limbs and ability to speak, swallow and breathe, whilst their mind and senses usually remain intact.
- The average life expectancy is 2.5 years from time of diagnosis.
- The mean time from onset of symptoms to confirmation of diagnosis is 10 to 18 months*
- It is estimated there are 2,094 Australians living with MND in 2015. Sixty per cent are male and 40% are female. The highest prevalence rate is reported in males aged between 75 and 84. The prevalence is estimated to be 8.7 per 100,000 Australians, or 1 in 11,434 Australians.

- Approximately 58% of people with MND are under the age of 65 and therefore are subject to different funding sources than those able to access aged care.

The Economic Impact of MND

The associated direct (e.g. health care, expensive equipment) and indirect (lost productivity and income) costs and intangible losses (independence, quality of life (QOL)) contribute to the high cost of this disease to society. According to the Deloitte Access Economics report (November 2015), "the total cost of MND is estimated to be $1.13 million per person with MND in 2015, total cost in Australia estimated to be $2.37 billion, productivity costs comprise 38% of the costs. The per-person costs of MND are substantially higher than a number of other diseases".

Notwithstanding the physical, psychological and emotional burden of the disease on MND family carers, the Deloitte report has quantified the economic disadvantage on families supporting people with MND who provide an estimate of 7.5 hours of informal care per day to people with MND: The productivity loss due to such informal care in Australia was estimated to be $68.5 million in 2015, or $32,728 per person with individuals shouldering most of these costs ($44.0 million), and with government bearing the rest ($24.5 million).

Healthcare Support for MND

MND remains a poorly diagnosed disease primarily because the symptoms are so varied and are inconsistent across generations. Many patients recall their very slow progression to diagnosis, often telling stories of having been sent to neurologists by their General Practitioners (GP's) with concern they have sustained a cerebral incident (haemorrhage, infarct [stroke] or trans-ischaemic attack [mini-stroke]) which has caused their peripheral limb weakness or slurred speech.

GPs across Western Australia now have access to a red flag diagnostic tool *Painless, progressive weakness – Could this be motor neurone disease?* The tool outlines MND signs and symptoms including bulbar and limb features, respiratory and cognitive features as well as supporting factors that point towards a diagnosis of MND. GPs and other health professionals can download the red flags diagnostic tool from the MNDcare website.

They also have access to an eight-page *Australian Doctor* article – *How to Treat Motor Neurone Disease* which is a comprehensive special feature and includes motor neurone disease case studies, signs, symptoms and 'red flags' for MND.

What has yet to be developed is a robust set of guidelines for GPs and neurologists in ensuring end of life (EOL) care commences for MND at the time of diagnosis (Aoun et al, 2016). Nor has a robust set of guidelines been crafted to explain in layman's language what EOL consists of, including palliative care.

Palliative Care

It is the international perspective and that of of MNDAWA that good palliative care commences from the time of diagnosis and encompasses symptom management, good information from multiple sources to meet each individual's learning style, and access to people who have the right information at the right time (Oliver and Aoun, 2013; NICE guidelines, 2016). Palliation for people living with MND is vastly more than ensuring access to narcotics and sedation at the end of life. Palliative care aims to optimise the quality of life of people with MND by relieving symptoms, providing emotional, psychological, and spiritual support to them and their

families, minimising barriers to a good death, and supporting the family post-bereavement. Therefore it is vital that people living with MND, their families and carers understand that palliative care is not just related to “the bit at the end”. For that reason, palliation for people living with MND is often progressive, like the disease, and often consists of removing interventions such as breathing or feeding assistance, rather than adding these as might be seen in other diseases such as cancer. For this reason, the approach used to care for people living with MND is quite broad and mutltidiscilinary.

MND Clinics

There are three specialised MND clinics in Western Australia:

1. The Perron Institute on the grounds of Sir Charles Gairdner Hospital with Dr Robert Edis as the lead neurologist:
2. The Fiona Stanley Hospital Neurophysiology Department with Dr Merrilee Needham leading; and
3. The St John of God Midland Public and Private Hospitals clinic led by Dr Lay Kun Kho.

Each of these clinics provide review and specialist medical and nursing staff to manage patients’ progression through their MND journey. People attending these clinics may be referred to respite, hospital and hospice care as is appropriate to their individual needs. Often discussions around Advance Health Directives (AHD) will be first approached by medical staff at these clinics. The nurses and Care Advisors (see MND Advisory Service below) who attend these meetings, are specialists who are able to provide advice and guidance about when and what to include in an AHD.

MND Advisory Service

MNDAWA is the pre-eminent specialist care and support organisation for people living with MND and provides care coordination and emotional support through the advisory service, which connects those living with MND to the services they require. The advisory service is also supportive of families and carers for people living with MND. Each patient registered with MNDAWA has a Care Advisor allocated to their support, enabling a close relationship to be established and the individual needs of each unique family situation to be supported. This includes encouraging families and carers to participate in education and support groups, equipment supply and hire, wheelchair accessible transport, specialized funding and patient advocacy. Findings from the association satisfaction surveys for the last three years show that the vast majority of patients and family carers find the MND Advisory Service of high value to them practically and emotionally, describing the service as “professional, supportive, always there, knowledgeable”.

The MND Advisory Service also provides access to, and advocacy for, people to complete their AHD, Enduring Guardianship documentation and Enduring Power of Attorney documentation. Associated with this service is a wealth of knowledge in providing sufficient information in a way that is easily understood, so people with the disease are able to make an informed decision about their AHD. Of equal importance, is the inclusion of families and carers in these discussions, so they know the decisions made are those of the person living with the disease and not something imposed on them.

AHDs may be as simplistic as a determination for not for cardiopulmonary resuscitation in the event of a cardiac or respiratory arrest, through to more complex directions such as whether or not to be fed via a percutaneous endoscopic gastrostomy (PEG) tube, or whether to receive narcotics or intravenous fluids.

Medications

The only treatment available is Riluzole, which has been shown to slow the rate of progression – but probably only by a few months – over a period of two years. However more recently, Edaravone has emerged as a drug with antioxidant properties. It protects nerve cells by mopping up damaging “free radicals” in the body. Research suggests the edaravone-treated group showed a significantly smaller decline in ALS Functional Rating Scale (ALSFRS-R) score compared with placebo. This suggests a beneficial effect of the drug over a period of 24 weeks in this cohort of people with ALS/MND. The group was not followed long enough to gather information on the longer-term effects and to establish impact on length of life. In 2015, regulatory bodies approved edaravone to treat people with MND in Japan and South Korea. MT Pharma America submitted an application to the FDA for regulatory approval in the US. This application was approved by the FDA on 5 May 2017. Mitsubishi Tanabe Japan is responsible for making an application to the Therapeutic Goods Administration (TGA) to approve edaravone for the treatment of MND in Australia. As yet no application has been made.

News stories on access to and use of medicinal cannabis often capture the interest of people living with MND. There are many online claims on treatments derived from cannabis that could slow progression or relieve symptoms for people living with MND; however, currently there is limited evidence or research supporting these claims.

Medicinal cannabis is cannabis and cannabinoids (usually tablet or oromucosal spray) that are prescribed by doctors for their patients. While Australia has recently legislated to allow for controlled cultivation of cannabis for medicinal and related scientific purposes, a viable industry is still some way off. In the meantime, legal cannabinoids can only be imported from a country where cultivation and manufacturing meet strict medicinal-grade standards.

Australia's regulatory authority for therapeutic goods, the Therapeutic Goods Administration (TGA) has not approved cannabis as a treatment for MND.

Respite

MND is a physically demanding disease for those providing care. In Western Australia, very few people with MND leave their home, relying upon unpaid carers, usually spouses or adult children, to provide the essentials of care, including mobilising, feeding and washing. These activities of daily living (ADL’s) become increasingly difficult as the person living with MND is less able to utilise their limbs.

Respite has always been available through various funding sources, but with the advent of the National Disability Insurance Scheme (NDIS) there have been some challenges in accessing these funds for people who have MND who are aged 65 years or over. Funds raised by volunteers and through donor appeals by MNDAWA are used to support respite when Federal or State health funding is difficult to obtain, or in the case of emergency.

Where respite used to be aimed at taking the person with the disease out of their home, the preferred method of respite is for a carer to be placed into the patient’s home, where they are in familiar surrounds and equipment. This change in direction over the past five years has seen an increase in people accessing respite and a significant reduction in short term hospitalisation or early hospice placement for people living with MND, especially toward the end of their life.

Equipment
Changes and developments in equipment used for people living with MND have also increased their longevity and quality of life. Advances in non-invasive ventilation and cough assist devices have enabled many patients to live at home until their death, as opposed to hospitalisation and inappropriate admissions to high dependency and even intensive care units.

The common use of media such as iPads and android equivalents has seen even older patients being able to communicate with their families and the wider community once their speech is impaired by failing motor neurones. Relationships with corporate enterprises to provide such devices at very low cost and then load write to speech software, has extended the quality of life for many and reduced repeat admission to Emergency Departments due to communication continuing. People living with MND know what is happening to them when things go wrong – with these communication devices, they can acquire the right assistance promptly. The advances in these communication software programs also enabled people living with MND to articulate their wishes in relation to how they wish their care to be managed, during their disease progression, and most importantly, at the end of their lives.

The Impact of the Disease on People with MND and their Family Carers
People living with MND do not have a mental illness. Their minds are as capable and aware as they were before their motor neurones (which affect muscle) started to fail. The complexity of the disease is that many people with MND have their hands and vocal cords affected by failing motor neurones, reducing their capacity to write and speak.

MND does not impact on any specific culture, nationality or social cohort more than any other. What is universal is the loss of control. How people come to terms with this loss of control varies depending on their innate personal qualities, their support systems, their financial situation and whether or not they have access to services (metropolitan versus country based). Along with this loss of control is the universal desire to be able to determine when they have "had enough". This mental line in the sand differs from person to person, from faith to faith and relates often to individual level of impairment.

People living with MND often speak/write of their frustration with not being able to do simple things like give their loved ones a hug *("my bloody arms don't work*"), enjoy a meal *("without drooling like a baby"*), and perform basic bodily functions unaided *("when your son has to hold your penis at the urinal, so you don't pee on your shoes, it forever changes your relationship*"). MND is a harsh, devastating and undignifying disease. Everyone with the disease eventually becomes dependent upon others to provide basic activities of daily living

The toll on family carers, particularly spouses who shoulder most of the care and support at home, has been widely described in the international literature and particularly from work in Western Australia (Aoun et al, 2012; Aoun et al 2017):

'I think you're traumatised, quite frankly. All the symptoms of trauma – numb[ness] and shock and all of those things'

"With cancer there is hope; with MND, there is nothing' If it had to be a choice between cancer and MND, I'd say cancer any day".

"They should bring in euthanasia - you wouldn't put a dog through what MND does - I find it very difficult. It really rips you apart."

"I mainly worry about how I'm going to cope. It's a terrible disease".

I do have to go to see "a shrink"— It's very stressful at times,"

"I turned around and I said, "Give me a kiss" and I thought at the time he was denying me a kiss, but I realise now he had no muscles in his face to kiss me with. I backed off and thought, it's not that you've lost feelings for me; it's just that you can't.

Many people with MND have to stop working and often their spouses also give up work to become a primary carer. This forces people to access financial support from the government or access superannuation savings, leaving little or nothing for the person who is left once they finally succumb to the disease. There is a perceived burden that is not only physical and psychological, there are massive financial burdens associated with this disease that are hidden from consideration when governments attempt to estimate the cost of the disease to the community.

Current Legislation

What does not change is their right to vote and their right to self-determination. As legislation and case law sits in Western Australia, this limits their capacity to merely declining (or refusing) hydration, nutrition, and essentially any intervention which may prevent death or prolong life. In 2009, a landmark decision was made by the Western Australian Supreme Court in favour of Mr Christian Rossiter, a paraplegic man living in a nursing home, to be able to decline nutrition through a PEG tube. While Mr Rossiter's situation was caused by an accident, his description of being a "prisoner in his own body" echoes the sentiments of people living with MND.

An AHD can be very prescriptive about what a person wants (or most often, refuses) in the management of their end of life care. Similarly, most Enduring Guardianship documents completed by people living with MND allowing a nominated person or persons, to make decisions regarding their healthcare, are about ensuring their life is not extended by clinical health intervention. Western Australian legislation for both the AHD and Enduring Guardian is very clear, but it does not permit the decision to be made to end a life. This remains outside legislation and is considered euthanasia and therefore murder or assisted suicide.

In its position statement, the peak body for MND, MND Australia, state that people living with MND have the right to:

"Accept, refuse, or discontinue treatment or intervention within the legal framework of the person's state or territory to ensure choice, control and the best quality of life possible, including access to preservation of personal dignity and to humane care, without discrimination".

Conclusion

There is a need for laws in Western Australia to allow citizens to make informed decisions regarding their own end of life choices. It is the position of MNDAWA that it is time for robust and considered discussion about the individual's right to self-determination and also their right to access appropriate medications, at a time of their choosing, to end their suffering with dignity and in a way, that is acceptable to each individual who has the disease, respecting their culture, religious faith and personal preference. We believe it is time to support their right to choose.

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