



**ENSURING SUSTAINABILITY OF CARE FOR  
THOSE WITH AMYOTROPHIC LATERAL  
SCLEROSIS (ALS) /MOTOR NEURONE  
DISEASE**

The Case for National Centre Funding

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## EXECUTIVE SUMMARY

Amyotrophic Lateral Sclerosis (ALS) also known as Motor Neuron Disease (MND) is a progressive neurodegenerative condition with a life expectancy of 3 years from first symptom. As diagnosis is often delayed by up to 15 months, most patients have less than 2 years of life expectancy from the time of diagnosis.

ALS fulfils the criteria for designation as a **Rare Disease**. The lifetime risk of developing ALS in Ireland is 1:300. The incidence is 2.7/100,000, which is close to that of Multiple Sclerosis. Approximately 140 patients are diagnosed with ALS in Ireland every year, and approximately 400 patients are living with the condition.

Evidence based guidelines from the American Academy of Neurology, the European Academy of Neurology and the UK NICE recommend that patients with ALS/MND should be managed by specialist multidisciplinary clinics.

In Ireland, this service is provided by the ALS/MND Group at Beaumont Hospital, which was the first centre in the world to show that patients have improved outcomes when treated in a multidisciplinary setting. The weekly clinic provides integrated care, with onsite specialist ALS/MND physicians, specialist health care professionals including neuropsychologists, experts in clinical measurement (respiratory function and functional assessment) and with representatives from the voluntary sector (**IMNDA**) as integrated members of the team. Patients receive coordinated care, with additional home-based outreach services including home visits provide by specialist nurses, psychologists and physicians.

**This is the only service of its kind in the country and multidisciplinary care is currently provided for over 80% of all ALS/MND patients living in Ireland.**

The Centre has embedded research into clinical practice, has an international reputation as a centre of excellence, and regularly receives referrals from other jurisdictions. For these reasons, The ALS /MND service at Beaumont Hospital has been recognized recently as a **National Centre of Expertise by the RCPI/HSE Rare Disease Programme** and has been endorsed by the HSE and Department of Health. Clinical staff have participated in the publication of a series of evidence-based guidelines for international use, provide ongoing in-service training, and have generated substantive guidelines for health care professionals. The service has also contributed to the HSE Neurology Programme Model of Care and has provided guidelines for best practice for management of ALS/MND in Ireland. **Research** is embedded into clinical practice and changes management (e.g. use of message banking for patients with evolving dysarthria; prospective studies of carer burden; validation of novel screening tools to assess cognitive and behavioural change in ALS; comparison of cough assists with breath stacking; evaluation and modification of staging protocols; development of a new scale to assess behavioural change in ALS; etc)

Despite its recognition as a National Centre by the Rare Disease Programme, the Centre has no designated ring-fenced funding, and most of the clinical staff are funded from research grants and philanthropic donations. Moreover, although patient care is coordinated in close collaboration with three specialist nurses from the Irish Motor Neurone Disease Association, the salaries for these their nurses is generated entirely by fundraising events. Equipment provided free of charge by the IMNDA is also purchased from fundraising, with limited support from exchequer funds.

**This document makes the case for supporting the internationally recognized high quality service for those with ALS/MND by providing sustainable funding for both the clinic, and the IMNDA nursing service, and the IMNDA equipment bank.**

## **BUDGET (See also Appendix, page 14)**

Support to enable “steady state” delivery of high quality care for all ALS/MND patients in Ireland is necessary. To achieve this, the following is required:

- Funding of outreach ALS/MND nurses by the HSE (currently supported by the Irish Motor Neuron Disease Association)  
€250,000 per annum
- Designation of an extra 0.5 WTE Consultant with expertise in ALS/MND  
€100,000 per annum
- Designated funding for 1.0WTE Specialist Training Fellow  
€90,000 per annum
- Designated funding to support the Irish ALS Register  
€75,000 per annum
- Designated funding to support 0.5 FTE Specialists in Physiotherapist, Occupational Therapist, Speech and Language Therapist and Nutritionists  
€200,000 per annum
- Designated funding for a Senior Grade Neuropsychologist  
€80,000 per annum
- Designated funding for a Senior Grade Social Worker  
€80,000 per annum

**Total Estimated Cost: €875,000 per annum**

## BACKGROUND AND DETAILED BUSINESS CASE

<p><b>Name of centre: Beaumont Hospital Amyotrophic Lateral Sclerosis/ Motor Neurone Disease Service</b>  <b>Address: Dept.Neurology,Beaumont Hospital. Dublin 09</b>  <b>Hospital CEO/Manager: Ian Carter</b>  <b>Hospital Group: Dublin NorthEast</b></p>	<p><b>Director of Service:</b>  <b>Professor Orla Hardiman , Consultant Neurologist, Beaumont Hospital</b>  <b>Professor of Neurology, Trinity College Dublin</b>          Email: <a href="mailto:Hardimao@tcd.ie">Hardimao@tcd.ie</a></p>												
<p><b>General Description of Condition and Service</b></p> <p>Amyotrophic Lateral Sclerosis(ALS) also known as Motor Neurone Disease (MND) is a rare neurodegenerative disease characterized by loss of all voluntary muscle activity, cognitive and behavioural change, and death within 3-5 years from first symptom.</p> <p>Approximately 140 new cases of ALS/MND are diagnosed in Ireland each year, and there are approximately 350 patients living with the condition.</p> <p>A national service for ALS/MND was initiated by Prof.Hardiman in 1994, with the support of the Irish Motor Neurone Disease Association (IMNDA). This service, which originated as a Consultant provided clinic, was developed as one of the first multi-disciplinary and cross-disciplinary services in the country, with extensive outreach programmes, teaching, training and research.</p> <p>The ALS/MND outpatient clinic now operates on a weekly basis, and currently engages with over 80% of all ALS/MND patients and their families in the Republic of Ireland. Patients are admitted to St. Brigids Neurology Ward in Beaumont Hospital for additional management where necessary, including nutritional assessment and gastrostomy insertion. However, the ethos of the service is to provide excellent care in an out-patient cross-disciplinary setting. This includes the integration of Hospital and Community based services, and extensive links with specialist Palliative Care.</p> <p>The ALS/MND Service prioritises management and care in the home. Respiratory management, including initiation and management of non-invasive ventilation (NIV) is undertaken in the home environment through the activities of a senior Clinical Nurse Manager, who provides this service for the entire country. The service also provides extensive psychological assessment and support for patient and caregivers, including initiatives for recognition and management of caregiver burden.</p> <p>Staff at the ALS/MND centre, in collaboration with the IMNDA provide regular lectures, seminars and workshops for healthcare professionals, caregiver and family members. The Centre has extensive research links to the Academic Unit of Neurology at Trinity College Dublin and the RCSI Clinical Research Centre.</p> <p>The service is recognized internationally as a Centre of Excellence, and the Centre is an active member of the European Network for the Cure of ALS (ENCALS, (of which Prof.Hardiman is Deputy Chair) and is a leading European Clinical Trial Centre for Phase II and Phase III trials of therapeutic agents for ALS/MND.</p>													
<p>Names of all medical consultants providing services at the centre and the WTE they each provide to this service.</p> <table border="0"> <tr> <td>Professor Orla Hardiman (Neurology)</td> <td>0.8 WTE (Director, and Principal Researcher)</td> </tr> <tr> <td>Dr.Margaret O’Brien (Neurology)</td> <td>0.2 WTE</td> </tr> <tr> <td>Dr.Niall Pender (Neuropsychology)</td> <td>0.2 WTE</td> </tr> <tr> <td>Dr.Richard Costelloe (Respiratory)</td> <td>0.1 WTE</td> </tr> <tr> <td>Dr.Regina McQuillan (Palliative Care)</td> <td>0.1 WTE</td> </tr> <tr> <td>Dr.Gerard Mullins (Neurophysiology)</td> <td>0.1 WTE</td> </tr> </table>		Professor Orla Hardiman (Neurology)	0.8 WTE (Director, and Principal Researcher)	Dr.Margaret O’Brien (Neurology)	0.2 WTE	Dr.Niall Pender (Neuropsychology)	0.2 WTE	Dr.Richard Costelloe (Respiratory)	0.1 WTE	Dr.Regina McQuillan (Palliative Care)	0.1 WTE	Dr.Gerard Mullins (Neurophysiology)	0.1 WTE
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<p><b>Voluntary Sector Involvement and Staffing</b></p> <p>The IMNDA works closely with the National ALS/MND services. The IMNDA funds 3 full time specialist nurses, who cover the entire country. These nurses attend the National Clinic, undertake home visits and facilitate integrated care across community had hospital-based services. The IMNDA also provides an equipment bank for patients, including mobility aids, communication aids and bedding, thus obviating the need for extending waiting periods for HSE-provided equipment, and reducing the cost burden for patients and their families.</p>													

The IMNDA staff liaise close with the staff provide services at the National ALS/MND clinic and facilitate expedited /emergency appointment for patients in need of urgent multi-disciplinary review. In turn, the Clinic accepts direct referrals from the IMNDA Nurses.

**Description of Total Activity**

<b>Number of:</b>	2014	2015	2016	2017
New consultations	140	145	180	200
Follow up consultations	1240	1300	1382	1430
Inpatient admissions	20	24	25	15
Average length of stay of inpatient admissions <b>(outliers not included)</b>	8 days	8 days	8 days	8 days
Day case consultations	0	0	20	24
Home visit consultations (approximation)	>400	>400	>500	>500
<b>Initiation of home based non-invasive ventilation</b>	33	35	40	50

**Current waiting time:**

- 1) a new consultation: less than 4 weeks
  - 2) follow up appointment: 1 week
  - 3) in-patient admission: 4 weeks
- Clinic operates an “open door” policy for emergency referrals and reviews

**Protected beds and emergency cover.**

While there are 12 in-patient Neurology beds on St.Brighids Ward, these are generally prioritise for admission through A+E. Patients requiring emergency admission for management of nutritional failure must wait on the Priority Waiting list, and unfortunately must occasionally seek admission through the A+E system .

This is not an ideal scenario, but a function of the current prioritization of A+E admissions over urgent “elective “ admissions. To facilitate excellence in the management of emergencies, a dedicated nurse- led phone service is available during work hours, and patients and caregivers can access the dedicated Neurology on-call service out of hours. The ethos of the Service is to ensure that patients with ALS/MND should not attend A+E, and to anticipate emergency by the nurse-led outreach service in collaboration with the nursing service provided by IMNDA.

In the increasing and distressing eventuality that an A+E attendance is required; the patients are reviewed by a member of the specialist ALS/MND team (during working hours) or by the Neurology on Call team (out of hours) and contact is made with the specialist team within 24 hours. Expedited access to Neurology beds is the preferred outcome, but not always feasible due to pressures within the Hospital system.

**Role of the Service in Providing Good Practice Guidelines**

- The service has participated in the publication of a series of evidence-based guidelines for international use (EFNS Guidelines published (2005, 2011, and latest version underway)
- The service has recently generated national guidelines for health care professionals for physiotherapy and speech and language therapy,
- The service has generated an evidence-based management algorithm for each stage of Motor Neurone Disease, which will shortly be available on the IMNDA website through the Health Professionals Portal
- The service has generated information booklets for Healthcare Professionals for management of Cognitive and Behavioural Change. A series of study days and workshops have been built around these.
- The services have generated information booklets for management of Cognitive and Behavioural Change for family members. In collaboration with the IMNDA, a series of workshops has been
- The service has contributed to the HSE Neurology Programme and has provided guidelines for best practice Model of Care for management of ALS/MND in Ireland.

- The Services is engaged internationally with initiatives to enhance the Patient Journey. This research component of the service is funded through various EU research funding programmes including Joint Programme in Neurodegeneration (ALS-CarE), and H2020 (TUDCA Clinical trial)

#### **Objective: A multi-disciplinary approach towards management of ALS/MND**

The ALS/MND service provides a week multidisciplinary clinic. This clinic provides integrated care, with onsite specialist ALS/MND physicians, specialist health care professionals (physiotherapy, occupational therapy, speech and language therapy, dietetics) and including neuropsychologists, clinical measurement (respiratory function and functional assessment); and with specialist nurse representatives from the voluntary sector as integrated members of the team.

A “patient passport” with details of the service and names of the clinical professionals both in the hospital and the community has been generated. This is provided to all patients and caregivers, as is an information sheet with details about the condition and useful numbers.

Further material, including guidelines for health care professional and caregivers for management of cognitive and behavioural impairment in ALS is also available through the clinic, as are materials provided by the IMNDA, the Irish Hospice Foundation and the Neurological Alliance of Ireland.

As noted, home based service to initiate non-invasive ventilation (NIV) has been in place since 2011. This specialist nurse –led service provides an opportunity to discuss end of life issues, implement advance care directives and incorporate educational secessions with community, hospital and hospice staff. In 2017, 50 patients were commenced on NIV at home. This represents a cost saving of over €150,000 to the hospital, (based on a requirement of a minimum of 3 days admission) and a saving of a minimum of 145 bed days. Details of the function of the multidisciplinary service have been published in over 60 peer reviewed publications specifically relating to MND care.

Evaluation of the clinic and comparison of outcomes between those attending the Multidisciplinary clinic and those who attend routine neurology clinics, or specialist clinics with devolved care (e.g. in Northern Ireland) has demonstrated superior outcome in survival for those attending the clinic at Beaumont Hospital (*Rooney et al, J Neurol Neurosurg Psychiatry. 2015 May;86(5):496-501*)

This service also provides a monthly dedicated clinic for post-polio syndrome in conjunction with the Irish post-polio support group, with on site and same day availability of specialist physiotherapy, and with expedited access to orthotics, rehabilitation services and specialist respiratory support.

#### **Evidence of capacity to adhere to good practice guidelines for diagnosis and care**

The ALS/MND service operates according to the European Federation of Neurological Sciences and the American Academy of Neurology practice guidelines, of both of which the Director (Hardiman) is an author. The service has embedded research into clinical practice and has implemented practice guidelines as part of an ongoing quality improvement cycle.

#### **quality management systems in place to assure quality of care, including National and European legal provisions**

Clinical intervention and outcome data are regularly reviewed and are subject to ongoing evaluation (e.g. stakeholder choice experiments; audit reviews etc). Management of “breaking sad news”, progression through the disease and approaching end of life decision making are compliant with published National and European guidelines.

#### **Participation in internal and external quality schemes, when applicable?**

The Service is part of a large European consortium that is currently assessing and evaluating the patient journey in ALS/MND, the use of various respiratory based measurements that increase the accuracy of outcome prediction and interventions (e.g. cough assist) that reduce intercurrent infections and enhance quality of life.

The service also undertakes regular self-audits of nursing and clinical professional activities.

#### **Capability to propose quality of care indicators and to implement outcome measures, including patient satisfaction initiatives.**

Quality of life measurement is embedded in to the services. Interventions are already subject to outcome analysis, for example:

- comparison of survival outcome and complication rates between PEG and RIG,
- comparison of vital capacity versus Sniff Nasal Inspiratory pressure as a predictor of survival
- Use of additional respiratory measures (peak cough flow) as survival predictors
- assessment of the beneficial effects of clinic- based shoulder injection by physiotherapists for

adhesive capsulitis in MND;

- comparison of breath stacking versus cough assist in management of secretions
- Evaluation of a novel cervical orthosis to support neck muscle weakness in neurological disease
- analysis of the benefits of voice banking
- use of mechanical insufflation /insufflation devices
- Effects of dysphagia and gastrostomy feeding on quality of life in ALS/MND
- Assessment of benefits of psychological intervention for caregiver burden (randomised trial)
- assessment of the benefit of exercise in post-polio syndrome (Randomised trial)
- Use of before and after quantitative muscle assessments to measure efficacy, optimal dose and timing of intravenous gamma globulin therapy for multifocal motor neuropathy.

Two longitudinal studies of the patient and carer journey in ALS/MND are also underway, funded by grants from the Health Research Board, the European Joint Programme in Neurodegeneration, The Irish Motor Neurone Disease Association and American ALS Association.

These studies incorporate quality of life measures for patient / carer dyads, carer burden studies, and choice experiments for patients and carers.

### **Evidence for an elevated level of expertise and experience**

- **Relevant publications from the Centre**

The ALS/MND Centre has generated over 250 peer reviewed publications over the past 12 years. These publications cover clinical aspects of ALS/MND including epidemiology, deep phenotyping, biomarker development, genetics, genomics, health services, health economics and drug development. The Centre also participates in the generation of publications for the ALS-Untangled programme

([www.alsuntangled.com/](http://www.alsuntangled.com/)) a website that examines the evidence for complimentary therapies in ALS.

- **Evidence of national/international recognition**

The ALS/MND Centre is a recognised ALS/MND centre of excellence by (1) the European ALS Consortium; (2) The American NEALS Consortium, and (3) by the World Federation of Neurology Subgroup on ALS/MND,

The Director of the MND Service (Prof.Hardiman) is Deputy Chair of the European Consortium and is recognised as an international opinion leader in ALS/MND. She is the Editor-in-Chief of the international peer reviewed journal *ALS the Frontotemporal Degenerations*; and is a member of the ENCALS Executive. She is also an advisor to industry and a leading international clinical trialist in ALS/MND.

The ALS/MND Centre has hosted the International Symposium on ALS/MND on 3 previous occasions (1995 ,2005, 2016). Over 1000 delegates from all over the world attend this meeting.

The Director (Hardiman) has been the Clinical Chair of each of these meetings and has been a member of the International Steering Committee.

The Centre Director is Chair of the Scientific Committee of the ENCALS meeting. She has provided numerous keynote addresses at international meetings on all clinical aspects of ALS/MND.

The Centre hosted the European ALS Consortium (ENCALS) meeting (over 300 delegates) in 2013 and 2015.

- **Involvement in production of national guidelines/care pathways**

The ALS/MND Group has authored the National Guidelines for management of ALS/MND, recently published by the Clinical Leads Programme in Neurology

### **Contribution to the scientific understanding of ALS /MND**

The Centre has published extensively in clinical care, deep phenotyping, biomarker development; epidemiology, genomics and health services related to ALS /MND (>220 peer reviewed publications in the past 10 years), and on outcome and rehabilitation in post-polio syndrome.

Specifically, research highlights include the following:

**2000: Arch Neurol** El Escorial Diagnostic criteria are not useful as prognostic indicators

**2003: JNNP Evidence** that multidisciplinary clinics are associated with improved survival in ALS

**2006 Nature Genetics** Discovery of ANG (ALS9) as causative gene in Amyotrophic Lateral Sclerosis

**2007 JNNP:** Survival of incident patients differ from prevalent patients with ALS

**2009 Neurology:** Conclusive evidence that rates of ALS are reduced in admixed populations

**2012 Lancet Neurology:** First description of detailed phenotype associated with C9orf72 repeat

**2012: Neurology:** Executive Impairment is a negative prognostic indicator in ALS

**2013: Ann Neurol:** Epidemiologic evidence of a genetic link between C9orf-72 and non-C9orf72 related ALS and neuropsychiatric disease

**2015: JNNP:** Centralized multidisciplinary clinical care is superior to devolved care in ALS

**2015: J Neurol:** Prognostic indicators for ALS can be identified on the first patient encounter

**2015: J Neurol:** Caregiver burden is increased by behavioural impairment in ALS

**2015: BMC Health SERV:** PR diagnostic patient journey is fragmented and poorly coordinated

**2016: Ann Clin Trans Neurol:** Evidence of behavioural heterogeneity in ALS

**2017: Nature Communications:** Genetic evidence of pathogenic overlap between ALS and Schizophrenia

**2017: Cerebral Cortex:** Evidence that EEG is a reliable marker of neurodegeneration in ALS

**2017: JAMA Neurol:** Neuropsychiatric endophenotypes aggregate within families

**2018: Neurology Genetics:** 30% of Irish ALS probands exhibit a family history of neurological impairment

**peer-reviewed publications**

>220 peer reviewed publications in the past 12 years.

**peer-reviewed grants supporting research**

In the past 8 years, the Centre has obtained over €10 million worth of competitive peer reviewed funding for research in ALS/MND. Grants within the past 5 years include the following:

ONGOING FUNDING				
Funding Body	Title of Proposal	Amount	StartDate	End Date
SFI	FUTURE NEURO (SFI CENTRE)	6 million (200K to the PI)	Feb 01 2018	Jan 30 2023
JPND	BRAINMEND	2 million €230 to the PI	Feb 01 2018	Jan 30 2021
SFI ERC Development	Deciphering ALS Heterogeneity	€480,000	01/09/16	30/08/2018
ALSA	A randomised control trial of psychological intervention for caregiver burden	\$200,000	01/01/18	31/12/19
ALSA	ALS Cognition: Profiling Caregiver burden	\$200,000	01/01/17	30/12/18
HRB (Dr. Bede's)	Emerging Leadership Award (Dr. Bede's) Imaging as a biomarker in ALS	€500,000	01/09/18	30/08/22
IRC	Spectral EEG as a biomarker in ALS (Roisin McMackin)	€100000	01/01/17	30/12/19
Science Foundation Ireland Partnership Award	Genome Sequencing of Irish ALS	€804,000	01/01/2016	30/12/2018
IICN Fellowship (Peter Bede)	Multimodal imaging in ALS: Analysis of mimic syndromes	€50,000	01/07/2016	30/06/20117
American ALS Association (ALSA)	The impact of cognitive and behavioural impairment on caregiver burden in ALS	\$200,000	01/04/2016	30/03/2018
MNDA Fellowship awarded to Dr. Russell Mc Laughlin	Whole-genome sequencing of 1000 Irish ALS patients and controls to identify novel ALS genes and pathways	€268,882	01/01/2015	30/12/2018
HRB Dissemination award	Cognitive and behavioural change in ALS; What caregivers need to know	€60,000	18/12/2015	17/12/2016
MNDA PhD Studentship	Language dysfunction in ALS	£120,000	01/07/2016	30/6/2019



Irish Research Council Bahman.	Analysis the motor networks in ALS and PLS	€100,000	01/10/2015	30/09/2017	
Research MN Rachel McConnell (Physiotherapy)	Mechanical Insufflation Exsufflation (M-IE) and Lung Volume Recruitment (LVR) in ALS/MND: a prospective study of the prescription process and patient outcomes.	€16,00	01/10/2012	30/09/2016	
Research MN Lesley Doyle(SLT)	Message banking and voice banking in ALS	€50,000	01/06/2015	30/5/2016	
Joint Programme in Neurodegeneration	ALS CarE (Coordinator)	2,000,000 Awarded	01/04/2014	30/03/2017	
Health Research Board (HRB) Fellowship Dr.Rooney	Environmental & Genetic epidemiology of ALS	€232,000	01/10/2014	30/09/2017	
IICN Fellowship (Tom Burke)	Social Cognition in ALS and Huntingtons Disease	€50,000	01/07/2014	30/06/2015	
Health Research Board (HRB) Clinician Scientist Award (Hardiman)	Deep Phenotyping and Complex Genetics of ALS: A multidisciplinary population based approach	€900,000	01/12/2012	30/11/2017	
EU FP7	EUROMOTOR (WP leader)	€230,000	01/01/2011	30/3/2016	
HRB (JPND)	Sampling and biomarker Optimization and Harmonization In ALS and other motor neuron	€232,000.	01/07/2012	30/06/2016	
IceBucket Award MND Association	Biomarkers, Deep Phenotyping and Genomics in ALS	€600,000	01/04/2015	30/03/2018	
Thierry Latran Foundation	Use of extended Irish kindreds to identify novel ALS associated variants	€100,000	01/07/2014	30/10/2015	
Project MinE	Genome Sequencing of Irish ALS	€800,000	01/01/2015	Ongoing	

Health Research Board (HRB)	Novel biomarkers of ALS subphenotypes using advanced imaging and spectral EEG technology	€329,772.00	01/10/2013	30/09/2016	
Health Research Board	Living and Dying with MND	€625,000.00	01/01/2013	30/12/2016	
Iris O'Brien Foundation	Building Leadership Capacity in ALS Research	€1,000,000	01/12/2014	30/11/2019	
ALS Association	Validating Staging in ALS	€75,000	01/07/2014	30/06/2017	
Elan Pharma Fellowship (Peter Bede)	Multimodal imaging in ALS	€240,000	01/07/2012	30/06/2017	

### Teaching and training activities

The Centre provides training for neurology house staff, health care professionals, nursing staff and psychologists.

This training takes place in 7 forms:

1. Attendance at the multidisciplinary clinic with debriefing and teaching sessions.
- 2.. Team meetings with research updates
3. Monthly supervision and de-briefing for junior staff on end of life issues and self-care
4. Formal workshops for peers within the hospital .
5. Presentation at journal clubs and clinical updates at monthly team meetings
6. Regular outreach teaching sessions, lectures and workshops for other health care professionals (averaging 1 -2 per month, delivered by different members of the team).
7. Formal lectures on the diagnosis and management of ALS/MND to students, peers and other healthcare professionals.
8. Annual day long multidisciplinary seminars for health care professionals .
- 9 .Engagement with the voluntary organization, with biannual meetings for patients. Yearly meetings with the Board of the organization.

### Provision of expert advice remotely

Email / telephone services are available through the hospital and using a dedicated website ([www.mnd.ie](http://www.mnd.ie) and [www.imnda.ie](http://www.imnda.ie) )

<p><b>Capacity to participate in data collection for clinical research and public health purposes</b></p> <p>A population based register has been in place for over 22 years, with complete ascertainment of all cases of MND and related conditions in Ireland .</p>
<p><b>Clinical trials</b></p> <p>Phase 2 and Phase 3 trials for disease modifying treatments in MND.</p> <p>Within the past 5 years, the Centre has conducted a large phase 3 trial (Dexpramipexole- Biogen -the highest recruiting site per capita in Europe); a phase 2 and Phase 3 trial for tirazemtiv-(Cytokinetics).,a Phase 2 study with Orion (LEVALS); a Phase 3 study with Cytokinetics (VITALITY), and a Methodology Trail with Biogen Idec. The Centre has been selected to participate n 5 further Phase 2 and 3 trials later in 2018</p> <p>A randomised controlled trial of the benefits of exercise in post polio syndrome (Investigator led)  Evaluation of a novel cervical orthosis to support neck muscle weakness in neurological disease (Investigator led in collaboration with Sheffield University.)</p>
<p><b>University Affiliation</b></p> <p>Trinity College Dublin, with additional links to the Royal College of Surgeons through the CRC.</p>
<p><b>Evidence of Multidisciplinary Approach</b></p> <p>The service provides an integrated approach with a “one stop shop” clinic through which patients receive care, with additional home based outreach services including home visits provide by specialist nurses, psychologists and physicians.</p> <p>In-clinic assessments include</p> <ul style="list-style-type: none"> <li>• Cognitive and behavioural screening</li> <li>• Functional assessment using standardised scales</li> <li>• Respiratory assessment (vital capacity and sniff nasal inspiratory assessment)</li> <li>• Measurement of transcutaneous blood gasses where indicted</li> <li>• training of patients in secretion management (breath stacking, use of cough assist)</li> <li>• management of shoulder pain using local injection</li> <li>• assessment of spasticity and suitability of treatment with botox</li> <li>• assessment of increased salivary gland secretion and suitability for treatment with botox</li> <li>• swallow studies</li> <li>• training in use of assistive communication devices</li> <li>• training and management of gastrostomies</li> <li>•</li> </ul> <p>Research is embedded into clinical practice and changes management</p> <p><i>In ALS/MND these include:</i></p> <ul style="list-style-type: none"> <li>• The use of message banking for patients with evolving dysarthria ; prospective studies of carer burden ;</li> <li>• validation of novel screening tools to assess cognitive and behavioural change in ALS;</li> <li>• comparison of cough assist (insufflation / exsufflation) with breath stacking;</li> <li>• evaluation and modification of staging protocols ;</li> <li>• development of a new scale to assess behavioural change in ALS;</li> <li>• assessment of different strategies to evaluate swallowing</li> </ul> <p><i>In post polio syndrome these include</i></p> <p>the use of quantitative muscle testing in assessing progression in post polio syndrome; the development of targeted exercise programmes and gait training in post polio syndrome, and the use of targeted quantitative measures to assess outcome following IVIg.)</p>
<p><b>systems in place to assure the continuity of care between all stages for each disease</b></p> <p>The disease is managed from diagnosis to end of life by the Centre, with strong links to the Palliative Care Services throughout the country.</p>
<p><b>Satellite/out-reach centres</b></p> <p>The service links with other Neurology centres in Ireland, and provides shared care for some patients who are not in a position to travel to the main clinic on a regular basis. The service also integrates seamlessly with the palliative care network of services throughout the country. Home visits are provided by the specialist nurse, clinical fellows and psychologists.</p>
<p><b>Links and collaborations with other centres of excellence at national, European and international level?</b></p>

The Centre is a member of the ENCALS Consortium ([www.ENCALS.eu](http://www.ENCALS.eu)) and the newly formed clinical trials network TRICALS . The Centre is also a member of the US based NEALS group.

#### **Links and collaborations with patient organisations**

The centre has extensive links with the Irish Motor Neurone Disease Association (IMNDA). The voluntary organization refers all newly diagnosed patients to the Centre. Patients attending the clinic are also automatically referred to the Association. An IMNDA nurse attends the weekly MDT clinic to maximise continuity of care for patients between both the Centre and the IMNDA. The Specialist Nurse based in the Centre works collaboratively with the three IMNDA nurses to provide a seamless outreach and home based service for all patients with ALS/MND and their families. Combined meetings are frequently organised with community services and palliative care services.

Members of the team speak annually at the AGM , and a joint meeting is held each summer entitled "Tea for MND". Each of these meetings is attended by large number of patients and carers amongst other stakeholders.

The IMNDA part funds a clinical fellow at the Centre, and is a partner on a HRB funded initiative to provide a series of countrywide educational talks to be delivered separately to health care professionals and patients/caregivers on the topic of cognitive/behavioural impairment in MND; how to recognise symptoms and strategies for managing behaviours.

#### **Innovations in the last 10 years to improve the delivery of care,.**

A detailed evaluation of the patient journey is underway, funded by the Health Research Board and the Joint Programme in Neurodegeneration. The initial findings of this work have been published in the following peer reviewed publications:

Galvin M, Madden C, Maguire S, Heverin M, Vajda A, Staines A, Hardiman O. Patient journey to a specialist amyotrophic lateral sclerosis multidisciplinary clinic: an exploratory study. BMC Health Serv Res. 2015 Dec 23;15:571.

Connolly S, Galvin M, Hardiman O. End-of-life management in patients with amyotrophic lateral sclerosis. Lancet Neurol. 2015 Apr;14(4):435-42.

Connolly S, Heslin C, and Mays I, Corr B, Normand C, Hardiman O. Health and social care cost of managing amyotrophic lateral sclerosis (ALS): an Irish perspective. Amyotroph Lateral Scler Frontotemporal Degener. 2015 Mar;16(1-2):58-62.

Rooney J, Byrne S, Heverin M, Tobin K, Dick A, Donaghy C, Hardiman O. A multidisciplinary clinic approach improves survival in ALS: a comparative study of ALS in Ireland and Northern Ireland. J Neurol Neurosurg Psychiatry. 2015 May;86(5):496-501.

Burke T, Elamin M, Galvin M, Hardiman O, Pender N. Caregiver burden in amyotrophic lateral sclerosis: a cross-sectional investigation of predictors. J Neurol. 2015 Jun;262(6):1526-32

Foley G, Timonen V, Hardiman O. "I hate being a burden": The patient Perspective on carer burden in amyotrophic lateral sclerosis. Amyotroph Lateral Scler Frontotemporal Degener. 2016 Feb 9:1-7. [Epub ahead of print] PubMed PMID: 26857752.

Foley G, Timonen V, Hardiman O. Acceptance and decision making in amyotrophic Lateral sclerosis from a life-course perspective. Qual Health Res. 2014 Jan;24(1):67-77.

#### **Examples of recently published studies relating to the delivery of care include the following:**

Elamin M, Bede P, Montuschi A, Pender N, Chio A, Hardiman O. Predicting prognosis in amyotrophic lateral sclerosis: a simple algorithm. J Neurol. 2015 Jun;262(6):1447-54.

Foley G, Timonen V, Hardiman O. Understanding psycho-social processes underpinning engagement with services in motor neurone disease: a qualitative study. Palliat Med. 2014 Apr;28(4):318-25.

Foley G, Timonen V, Hardiman O. Exerting control and adapting to loss in amyotrophic lateral sclerosis.

Soc Sci Med. 2014 Jan;101:113-9.

Murray D, Hardiman O, Meldrum D. Assessment of subjective and motor fatigue in Polio survivors, attending a Post polio clinic, comparison with healthy controls And an exploration of clinical correlates. *Physiotherapy Theory Pract.* 2014 May;30(4):229-35. Doi: 10.3109/09593985.2013.862890. Epub 2013 Dec 16. PubMed PMID: 24328931.

Elamin M, Bede P, Byrne S, Jordan N, Gallagher L, Wynne B, O'Brien C, Phukan J, Lynch C, Pender N, Hardiman O. Cognitive changes predict functional decline in ALS: a population-based longitudinal study. *Neurology.* 2013 Apr 23;80(17):1590-7.

Murray D, Meldrum D, Moloney R, Campion A, Horgan F, Hardiman O. The effects of a home-based arm ergometer exercise programme on physical fitness, fatigue and activity in polio survivors: protocol for a randomised controlled trial. *BMC Neurol.* 2012 Dec 13;12:157.

Hardiman O. Multidisciplinary care in ALS: expanding the team. *Amyotroph Lateral Scler.* 2012 Feb;13(2):165. doi:

Bede P, Oliver D, Stodart J, van den Berg L, Simmons Z, Ó Brannagáin D, Borasio GD, Hardiman O. Palliative care in amyotrophic lateral sclerosis: a review of current international guidelines and initiatives. *BMJ Support Palliat Care.* 2011 Dec;1(3):343-8.

Foley G, Timonen V, Hardiman O. Experience of services as a key outcome in amyotrophic lateral sclerosis (ALS) care: the case for a better understanding of patient experiences. *Am J Hosp Palliat Care.* 2012 Aug;29(5):362-7.

Bede P, Oliver D, Stodart J, van den Berg L, Simmons Z, O Brannagáin D, Borasio GD, Hardiman O. Palliative care in amyotrophic lateral sclerosis: a review of current international guidelines and initiatives. *J Neurol Neurosurg Psychiatry.* 2011 Apr;82(4):413-8.

**CURRENT WTE STAFFING**

Staff member	Number of WTE (available to the MND Service)
Consultant	1.2
NCHD	0
Specialist Registrars	0
CNS/ANP	1.0
Staff Nurse	0.5 (Day Unit)
Allied Health Professionals:	Specialists in Physiotherapy, Occupational Therapy, Speech & Language Therapy, Dietetics attend the clinic. However, they are also charged with other duties across the hospital and have limited availability outside clinic hours.
Medical Social Worker	No social worker allocation to the services
Clinical Psychologist	5 (research assistants (funded by grant income) (Director of Neuropsychology is actively involved but is also responsible for a wide range of services across the hospital

Business Manager	The Clinic has access to a Business Manager Neurology Directorate at Beaumont Hospital The Academic Unit at TCD (providing research support) has as Business Manager
Other : Research Registrars (5 -funded from grants and philanthropy) Clinical Trial Nurses (x2) & managers Research Manager (x1 funded from grant income) ) Research Team (x 20 funded from grant income).	
<p><b>Service Cost Savings Initiatives</b></p> <p><b>Health Economics</b> A health economic study is currently underway to identify the cost of diagnosis; to identify means by which diagnoses can be streamlined and diagnostic delay reduced; and to determine the overall costs (direct and indirect ) of ALS/MND from first symptom to death.</p> <p><b>NIV</b> An out-reach service was commenced to introduce non-Invasive Ventilation at home reducing hospitalization for patients. In 2015, 45 patients were commenced in NIV at home, leading to a saving of over 150 bed days. Monitoring of oxygen saturation is undertaken at 3 month intervals using remote technologies, obviating the need for 3 monthly overnight saturating monitoring and providing further cost savings.</p> <p><b>Tailoring Treatment to Patient Needs</b> IVIg therapy for patients with multifocal motor neuropathy is tailored to the patient’s needs using quantitative muscle testing. Treatment is provided in the Neurology Day Unit. A programme of home treatment has been initiative for some patients, reducing the costs of admission.</p> <p><b>Awareness of Cognitive and Behavioural Change</b> Our newly established programme of training for health care professionals in dealing with behavioural difficulties in ALS has the potential to reduce the number of emergency contacts, as the training builds expertise in the community</p>	
<p><b>UNMET NEEDS AND FUNDING REQUIREMENTS</b></p> <ol style="list-style-type: none"> <li>1. Funding for a second specialist ALS/MND nurse based in the Centre</li> <li>2. Funding for 0.5WTE consultant with a specialist interest in ALS/MND, and a desire to continue as an international figure in ALS/MND management and research.</li> <li>3. Protected funding for clinical professionals (0.5WTE each in PT, OT, SLT, Dietician)</li> <li>4. Protected funding for a dedicated Neuropsychologist</li> <li>5. Protected funding for a 0.5 dedicated Social Worker</li> <li>6. Exchequer funding for 3 WTE Outreach Nursing services (currently funded through IMNDA)</li> <li>7. Upgrade of Poorly serviced clinic waiting area and consultation rooms for individuals with disabilities e.g. limited space for wheelchairs, inappropriate seating.</li> <li>8. Transport for patients travelling long distances.</li> <li>9. Sustainable funding for the Irish ALS/MND Register</li> </ol>	
<p><b>Key recommendations to improve delivery of patient centred care for Centre (from the RCPI/HSE Rare Disease Assessment process)</b></p> <ol style="list-style-type: none"> <li>1. Overall development of the Centre with a ring fenced budget to ensure sustainability over the coming 10 years, following retirement of the current Director.</li> <li>2. Sustainable integration of IMNDA services with National Centre, including sustainable funding of IMNDA nurses</li> <li>3. Funding for the ALS/MND Registers (Longest running and most complete Register of its kind in the world)</li> </ol>	
<p><b>APPENDIX : VISION AND BUDGET</b></p>	

The ALS/MND Centre based at Beaumont Hospital, with academic links to Trinity College Dublin is a world leader in epidemiology, clinical phenotyping, neuropsychology, imaging, genomics, and clinical trials.

The core aspects of the Centre are funded through the Hospital – however the service is under-resourced and is reliant on a cohort of highly dedicated and motivated staff. Research is supported by competitive peer reviewed grant funding.

Clinical trials are undertaken by a dedicated specialist team, funded on an ongoing basis from clinical trial overheads.

The ALS Register is the longest running register of its kind in the world. It is funded from a combination of grant funding and research overheads.

To ensure the continued sustainability of the excellent service and to enable further development at both national and international levels, its designation as a National Centre of Excellence for ALS/MND with a commensurate budget is required. The Centre has already been designed by the Rare Disease as a National Centre of Excellence, and eligible for consideration as a European Reference Centre. However, no funding has been attached to the centre.

Support to enable “steady state” delivery of high quality care for all ALS/MND patients in Ireland is necessary. To achieve this, the following is proposed:

### **1. Funding of outreach ALS/MND nurses by the HSE in association with the Irish Motor Neuron Disease Association**

**Rationale:** The Irish MND Association currently provides funding for 3 nurses who provide outreach /liaison services for ALS/MND patients. The salaries for these nurses are currently supported exclusively by fundraising within the voluntary sector and are at constant risk. A similar population (e.g. Scotland) is supported by 16 WTE specialist nurses, funded by the Scottish Health System.

Provision of funding for these 3 WTE nurses will embed the current essential outreach service within the Health Sector through the Centre of Expertise and will ensure uninterrupted high quality integrated care for patients with ALS/MND.

There is strong evidence that provision of such a service reduce hospital admissions and limits the use of emergency interventions including invasive ventilation.

**Total Cost : €250,000 per annum**

### **2. Designation of an extra 0.5 WTE Consultant with expertise in ALS/MND**

**Rationale:** Additional consultant support is required to provide a consistent consultant-provided service. This has been recommended by the Rare Disease Office (endorsed by the Dept. Health) in the context of the European Rare Disease Network. As ALS is a rare disease, specific expertise is required to ensure quality, and to introduce and disseminate advances in best practice.

The provision of an extra 0.5WTE post also ensures that the quality ALS/MND services are retained following retirement of the incumbent clinician.

**Total Cost: €100,000 per annum**

### **3. Designated funding for 1.0WTE Specialist Training Fellow**

**Rationale:** The delivery of care for a rare disease such as ALS/MND, (which can also be viewed as a model of neurodegeneration in general) requires ongoing education of junior staff.

This post will ensure ongoing knowledge and skills transfer to enable long term excellence in continuity of care.

**Total Cost : €90,000 per annum**

### **4. Designated funding to support the Irish ALS Register**

This has been recommended by the Rare Disease Office.

**Rationale:** The value of population-based registers is increasingly recognized by the Health Services. Not only can population-based registers provide detailed and fine-grained data regarding disease enabling data-drive health care planning, the Irish ALS Register has also facilitated significant world class research in clinical care, deep phenotyping, biomarker development; epidemiology, genomics and health services

related to ALS /MND (>200 publications in the past 10 years).

**Total Cost: €75,000 per annum**

**5. Designated funding to support 0.5 FTE Specialists in Physiotherapist, Occupational Therapist, Speech and Language Therapist and Nutritionist**

**Rationale:** There is considerable evidence that intensive specialist health care professional intervention in ALS/MND improves respiratory symptoms, reduces falls, enhances communication, reduces aspiration pneumonia, reduces hospitalization and improves quality of life.

**Cost: (€200,000 per annum**

**6. Designated funding for a Senior Grade Neuropsychologist**

Up to 50% of patients with ALS develop cognitive and behavioural change. Detailed assessment is required to establish the level of impairment. Behavioural change also leads to significant caregiver burden, and neuropsychological counselling supports are required.

**Cost: €80,000 per annum**

**7. Designated funding for a Senior Grade Social Worker**

ALS/MND affects the entire family, and the wider community. Patients and their families must negotiate a wide range of services. Determining entitlements can be both complex and stressful. As up to 50% of people who develop ALS are still working, and up to 30% still have dependent children, the financial implications can be considerable. Many patients and the families require a designated social worker to help negotiate the process of care provision.

**Cost: €80,000 per annum**

