

D04/S/a

**2013/14 NHS STANDARD CONTRACT
FOR NEUROSCIENCES: SPECIALISED NEUROLOGY (ADULT)**

SCHEDULE 2 – THE SERVICES – A. - SERVICE SPECIFICATIONS

Service Specification No.	D04/S/a
Service	Neurosciences: Specialised Neurology (Adult)
Commissioner Lead	
Provider Lead	
Period	12 months
Date of Review	

1. Population Needs

1.1 National/local context and evidence base

A specialised neurology service will provide people who have a complex single need or multiple conditions with responsive specialist services using multi-disciplinary teams and disease-specific protocols and pathways, such as those specified in the relevant National Service Frameworks and Quality and Outcomes Framework. Pro-active care management can make a real difference to patients with a single condition or a range of problems that threaten their health and well-being and can produce better health outcomes, slow disease progression, reduce disability, ensure better management of sudden deteriorations often associated with long term conditions and result in improved quality of life for patients, as set out within the NHS Outcomes Framework 2012/13, and reduced need for admissions to hospitals.

2. Scope

2.1 Aims and objectives of service

Aims

Specialised Neurology Services will:

- Improve clinical specific outcomes
- Improve the quality of Neurology services and patient experience.
- Reduce the number of unscheduled admissions and re-admissions to hospital with neurological conditions.
- Reduce hospital length of stay, and thus occupied bed days.

- Improve referral and signposting to appropriate services for patients, including voluntary agencies and community groups.
- Improve transitional care.
- Increase patient choice.
- Reduce inequalities in health between those with neurology condition, both across the region and with other parts of the United Kingdom.
- Facilitate future research into these conditions, through “academic neurology”, research led by specialist nurses and other health professionals, and research into best ways of providing service.
- Facilitate training of future consultants neurologists in these conditions, and of specialist nurses and allied health professionals essential for the care of these patients.

Objectives

- Provide improved quality of life and patient experience, for patients and for their carers.
- Provide a service that is accessible for individuals and their families, ensuring standardisation of care across the specialist centres.
- Offer the best services within the resources available.
- Provide high quality care for all individuals with smooth transitional care for young adults and effective support for families.
- Provide care as close to homes with regular review by local service providers, specialist centres act as coordinator to ensure appropriate care plans are in place to ensure lines of communication and whole system management of patients.
- Specialised services will be available within each NHS region to support local provision where appropriate.
- Meet national standards and constantly audit the service to document that the best possible outcomes and patient experience are delivered.
- Ensure a patient-centred approach to the delivery of services.
- Improve access to services in primary care through referral to more integrated services, particularly for disadvantaged groups and geographical areas.
- Encourage a range of approaches for case- management.
- Encourage greater participation in self-care through appropriate support mechanisms.
- Improve the availability of clinical and non-clinical information.
- Use innovative technologies to enable those living remote from neurology centres to receive the same care as those living close by.
- Enable advancement of future care through research and enhanced training.

2.2 Service description/care pathway

Service Description

A neurology service should be regarded as specialised when a patient is referred from a consultant or a professional with specialist training in the condition to a consultant neurologist with sub specialist expertise for

investigation or treatment that requires the special expertise that is contained within that pathway of care that lies out with the expertise of the referring consultant. This would include that referrals such as those to a neurologist with a special interest in neuro-ophthalmology by a consultant ophthalmologist (i.e. a specialty out with neurology). Furthermore, referral from a physiotherapist assessing patients in a musculoskeletal service to a consultant neurologist in the Multiple Sclerosis (MS) clinic would be included. Neither of these are uncommon and represent ways in which patients “find their way” to subspecialist clinics.

AND/OR

Where best practice e.g. NICE guidance, would require multi-professional care that might include involvement of **more than one** other professional group within the defined service e.g. specialist nurse, allied health professionals, orthotists, dieticians, speech and language therapists, psychologists or psychotherapists, continence services, pain relief services and respiratory care services.

AND/OR

Where there are joint arrangements with specialists in rehabilitation medicine, cardiology and clinical genetics, ophthalmology, otology, interventional radiology, neurosurgery, orthopaedic and spinal surgery where their involvement is an integral part of patient care e.g. patients with neuromuscular disorders, ataxia. This would include access to sleep studies where clinically appropriate.

AND/OR

As part of care where the patient's condition is such that it involves assessment for potential elective neurosurgical procedures or interventional radiography, (e.g. patients with epilepsy who have seizures resistant to medical treatment, or those with complex movement disorders,) and their subsequent follow up after surgery.

AND/OR

As part of care that involves drugs for which special funding or expertise is required e.g. botulinum toxin for dystonia or spasticity management, apomorphine for Parkinson's Disease (PD), intravenous immunoglobulin for accepted neurological indications, disease modifying therapies in MS and other neuro-inflammatory disorders, recently licensed drugs for epilepsy and drugs on the current high cost drugs list.

AND/OR

Where care is part of transitional arrangements between child and adult services for adolescents with multiple comorbidities (in addition to a primary neurological diagnosis) e.g. children with epilepsy or movement disorders who have learning disabilities, physical handicap or other non-neurological comorbidities or those with a severe isolated single neurological problem .

Sub-Specialities

Inflammatory Disorders of the Nervous System

Services should be provided across all ages supporting multidisciplinary care for people with multiple sclerosis, neuromyelitis optica and other rare inflammatory disorders of the Central Nervous System (CNS) as set out in National Institute for Health and Clinical Excellence (NICE) guidelines and including International Classification of Diseases (ICD) codes G35 – G37.

Specialised services for inflammatory disorders of the nervous system will include; multi-professional care including involvement of: specialist nurses, professions allied to medicine, orthotists, dieticians and speech and language therapists, psychologists, continence and pain relief services, services provided jointly with specialists in rehabilitation medicine, spasticity management services and clinics for the assessment for and monitoring of disease modifying therapies.

Epilepsy and related disorders

Specialised services for complex or intractable epilepsy (ICD G40) will include: multi-professional care including involvement of: epilepsy nurse specialists, clinical psychologists, dieticians and learning disability services, pre-surgical assessment services, clinics providing care for those with seizures resistant to treatment and arrangements for transitional care between paediatric and adult clinicians.

Movement disorders

Including e.g. Parkinson's disease, Progressive supranuclear palsy, Steele Richardson Syndrome, Striatonogral degeneration, dystonia and other movement disorders (G20 – G26 and G90).

Specialised services will include those clinics providing multi-professional care might include involvement of: Parkinson's disease nurses, allied health professionals, dieticians and speech and language therapists, psychologists. Pre-surgical assessment services, neurogenetics and botulinum toxin clinics and those providing transitional care between paediatric and adult clinicians.

Neuromuscular Conditions

Specialised neuromuscular services will be provided to neuromuscular disorders (NMD) patients of all ages and via a managed clinical pathway that supports multi-disciplinary and cross organisational working. This will facilitate effective and efficient treatment care and support to patients, their families and carers.

There are more than 60 different types of muscular dystrophy and related (NMD). Neuromuscular disorders can be genetic or acquired:

Inherited neuromuscular disorders include:

- Muscular dystrophies

- Spinal muscular atrophy (SMA)
- Congenital and syndromal neuropathies
- Inherited neuropathies
- Congenital myopathies
- Metabolic myopathies
- Genetic myasthenic syndromes
- Mitochondrial disorders
- Channelopathies
- Myotonias

Acquired disorders include:

- Myasthenia gravis
- Autoimmune neuropathies
- Inflammatory myopathies.

There are few curative treatment options for most of these diseases. A number, such as Duchenne Muscular Dystrophy, are aggressive and cause progressive muscle wasting and weakness, orthopaedic deformity, cardiac and respiratory compromise, dependency on others for day-to-day care and usually result in premature death. Others (e.g. Hereditary Motor and Sensory Neuropathy) cause life-long disability without limiting life span.

Some neuromuscular disorders can present in childhood (e.g. Duchenne Muscular Dystrophy) or young adult life. Others can be late onset conditions in adulthood (e.g. inclusion body myositis)

The service specification for neuromuscular conditions developed as part of the national work programme is annexed to this document as an example of good practice (Annex B)

Stroke

Specialised stroke services will only include the investigation of patients with stroke of non athero-embolic origin (carotid dissection, stroke of cardiac origin, stroke due to coagulation disorders), patients with genetically determined stroke, Arteriovenous malformation (AVM) and cerebral vasculopathies such as vasculitis. Clinics providing transcranial doppler examinations and joint services with cardiology including trans-oesophageal echocardiography (TOE). All other stroke activity will be commissioned by Clinical Commissioning Groups.

Motor Neurone Diseases (MND)

The description of a specialised motor neurone disease service is where care for a patient with motor neurone disease involves a multi-disciplinary approach from MND specialists, including a specialist neurologist, to direct care and provide diagnostic certainty, a specialist nurse / care co-ordinator and specialist provision in respect of: respiratory support, speech and language therapy, dietetics and nutrition, physiotherapy, saliva management, respiratory secretion management,

neurorehabilitation, occupational therapy, gastroenterology and gastrostomy, chiropody, management of cognitive impairment, riluzole safety blood tests, palliative care, end of life care, counselling / emotional support, telephone support.

It is important to note that MND is among the most demanding neurological conditions. It is degenerative, often rapidly so, and requires complex and anticipatory care significantly above what a general neurology service can provide.

People with MND will, in varying sequences and combinations, lose the ability to speak, swallow and use their limbs; the most common cause of death is respiratory failure. Most commonly the individual will remain mentally alert as they become trapped within a failing body, although some experience dementia or cognitive change. There are about 5,000 people living with MND in the UK. Half of people with the disease die within 14 months of diagnosis.

All services for people with motor neurone disease should be commissioned as a specialised service

Neurogenetics Diseases

Neurogenetic services are provided through a partnership between neurology centres and specialist genetic centres. Clinical counselling and predictive genetic testing for neurological disorders that have a genetic or familial association are included in this definition where this activity takes place within a neurosciences or neurology only centre.

Neuro-Oncological Diseases

Includes patients with primary and secondary brain tumour and those with the remote effects of cancer on the nervous system. Specialised services for neuro-oncology may include: multi-professional care including joint involvement of: oncology specialist nurses, professions allied to medicine, dieticians, clinical psychology, speech and language therapists. Joint oncology clinics including those for low grade and high grade glioma patients, services for patients with the remote effects of cancer and joint palliative care clinics.

Specialist Clinic for Cognitive Disorders

Includes patients presenting with memory and other disorders of cognition, e.g. Alzheimers disease, non Alzheimer dementias (frontal temporal dementia), Creutzfeldt Jacob Disease, dementias of vascular and metabolic origin. Specialised services for cognitive disorders should include: diagnostic services for younger patients (under 65 years) and inherited dementias, joint therapy services with mental health services teams and joint working with neuropsychology services, and with neurosurgeons, and include patients in whom the cognitive disorder is post neurosurgical.

Service Model: Generic Service Model

Specialist services will operate at three levels:

Tier 1: Local community and primary care services e.g. physiotherapy and community nursing where skills can be developed with the assistance of specialist staff from the Hub centres in order to provide ongoing basic maintenance for people with neurological conditions

Tier 2: Multidisciplinary specialist outreach clinics to be developed in main population centres. These outreach services, provided by the members of the multidisciplinary team visiting from the hub centres, will constitute the “subspecialist spokes”

Tier 3: Specialist Care provided at a tertiary centre acting as a ‘Hub’ and fully equipped to carry out the full range of neuro-related procedures, investigations and treatments such as muscle biopsies and early access to other neuromuscular specialist clinical services such as respiratory, cardiac and orthopaedic.

This model would be supported by the following:

- A ‘virtual’ Clinical Network - to support multidisciplinary and cross organisational working to provide effective and efficient treatment, care and support to patients and their families.
- Shared care with services delivered as close to home as possible as well as access to a specialist centre, supported by multidisciplinary team working - each person will be supported by an individualised package of care.
- Availability of support within each region and will offer emotional support, care co-ordination and to translate medical information into a format families can understand
- Individualised care supported by ongoing care coordination from the point of diagnosis.
- Close working between paediatric and adult services to ensure smooth transition between services

2.3 Population covered

The service outlined in this specification is for patients ordinarily resident in England*; or otherwise the commissioning responsibility of the NHS in England (as defined in *Who Pays?: Establishing the responsible commissioner* and other Department of Health guidance relating to patients entitled to NHS care or exempt from charges).

* - Note: for the purposes of commissioning health services, this EXCLUDES patients who, whilst resident in England, are registered with a General Practitioner (GP) Practice in Wales, but INCLUDES patients resident in Wales who are registered with a GP Practice in England.

The population served should be covered by the named neuroscience centres (neurosurgery and neurology) Included in this specification are those whose needs are outlined in 2.2 above.

2.4 Any acceptance and exclusion criteria Location(s) of Service Delivery

- Each region will have a directory of services that will highlight:
- Specialist adult and children's neurology specialised services available within the Region
- Specialist neurology centres that take referrals from the main children's and adult units
- Availability and location of specialist Neurology Rehabilitation Services
- The existing model of neurology and neurology /neurosurgery centres should be preserved and neurologists should not be appointed unless they have contracts that include as a minimum 2 Programmed Activities (PA's) for continuing professional development and governance at such a centre

Referral Route

Between a consultant or a professional with specialist training in the condition to a consultant neurologist with sub specialist expertise for investigation or treatment that requires the special expertise that is contained within that pathway of care that lies out with the expertise of the referring consultant.

Discharge Criteria and Planning

Many patients will have long term, deteriorating conditions where discharge is rare. If patients feel they are sufficiently supported locally and are stable, they will be given open appointments for them to be reviewed as needed and every patient requiring specialised follow up must be given an emergency care plan, including how to access the specialist service

2.5 Interdependencies with other services

The following services are integral to the care of people with neurological conditions and the commissioning of specialised neurology services should integrate with the commissioning arrangements for neurology / neurosurgery intensive care.

- Neurosurgery
- Neuroradiology
- Neurophysiology
- Neuropsychology
- Neuropsychiatry
- Neuropathology
- Neurological rehabilitation
- Specialised Equipment Services

Service Specifications for the following services should also be considered:

- Neurosurgery
- Neuro rehabilitation
- Complex Disability Equipment

3. Applicable Service Standards

3.1 Applicable national standards e.g. NICE, Royal College

NICE Clinical Guidelines on Epilepsy

NICE Clinical Guidelines for MS

NICE Guidelines on Deep Brain

Stimulation NICE Guidelines for

Parkinson's Disease

Also

Movement Disorder Society (MDS) and Draft Neuromuscular Service Specification

NICE Quality Standards Epilepsy (in production).

Quality, Innovation, Productivity and Prevention (QIPP) Accountable Integrated Care

Systems right care workstream developing epilepsy and Parkinson's and MS template

Long term conditions NSF and new Long Term Conditions Outcomes Strategy
National Strategy for Stroke

Royal College of Physicians (RCP) Guidelines and Reports including Local
Adult Neurology Services for the next decade

<http://bookshop.rcplondon.ac.uk/details.aspx?e=354>

4. Key Service Outcomes

- Improve the quality of patient experience for all patients with a neurological condition experience
- Reduce the number of unplanned admissions and re-admissions to hospital
- Reduce hospital length of stay
- Compliance with national access and time to treatment targets
- Reduce inequalities
- Meet NICE guidelines

- Improve referral and signposting to appropriate services for patients
- Improve transitional care through greater integration between paediatric and adult care services.

Preventing people from dying prematurely through:

- Ensuring patients are monitored regularly to allow for early identification of changes in their disease progression
- Development of emergency care plans for all patients with a long term neurological condition

Enhancing Quality of Life for People with Long Term Conditions through:

- Agreeing individualised care plans for all patients with long term conditions
- Sign posting patients to appropriate services and equipment services

Helping people to recover from episodes of ill health or following injury through:

- Reducing 30 day readmission rates
- Ensuring referral to appropriate nurse specialists

Ensuring that people have a positive experience of care through:

- Monitoring Complaints
- National Outpatient Survey
- National Inpatient Survey

Treating and caring for people in a safe environment and protecting them from avoidable harm through:

- Venous thromboembolism (VTE) risk assessments
- Monitoring prescribing errors

Annex A Neurosciences Centres (Source: Identification Rule)

RF4	BARKING, HAVERING AND REDBRIDGE UNIVERSITY HOSPITALS NHS TRUST
RNJ	BARTS AND THE LONDON NHS TRUST
RXH	BRIGHTON AND SUSSEX UNIVERSITY HOSPITALS NHS TRUST
RGT	CAMBRIDGE UNIVERSITY HOSPITALS NHS FOUNDATION TRUST
RWA	HULL AND EAST YORKSHIRE HOSPITALS NHS TRUST
RYJ	IMPERIAL COLLEGE HEALTHCARE NHS TRUST
RJZ	KING'S COLLEGE HOSPITAL NHS FOUNDATION TRUST
RXN	LANCASHIRE TEACHING HOSPITALS NHS FOUNDATION TRUST
RR8	LEEDS TEACHING HOSPITALS NHS TRUST
RVJ	NORTH BRISTOL NHS TRUST
RX1	NOTTINGHAM UNIVERSITY HOSPITALS NHS TRUST
RTH	OXFORD RADCLIFFE HOSPITALS NHS TRUST
RK9	PLYMOUTH HOSPITALS NHS TRUST
RAL	ROYAL FREE HAMPSTEAD NHS TRUST
RM3	SALFORD ROYAL NHS FOUNDATION TRUST
RHQ	SHEFFIELD TEACHING HOSPITALS NHS FOUNDATION TRUST
RTR	SOUTH TEES HOSPITALS NHS FOUNDATION TRUST
RHM	SOUTHAMPTON UNIVERSITY HOSPITALS NHS TRUST
RJ7	ST GEORGE'S HEALTHCARE NHS TRUST
RTD	THE NEWCASTLE UPON TYNE HOSPITALS NHS FOUNDATION TRUST
RET	THE WALTON CENTRE NHS FOUNDATION TRUST
RRV	UNIVERSITY COLLEGE LONDON HOSPITALS NHS FOUNDATION TRUST
RRK	UNIVERSITY HOSPITAL BIRMINGHAM NHS FOUNDATION TRUST
RJE	UNIVERSITY HOSPITAL OF NORTH STAFFORDSHIRE NHS TRUST
RKB	UNIVERSITY HOSPITALS COVENTRY AND WARWICKSHIRE NHS TRUST

Annex B

Shared outcomes

Specialised Neuromuscular Services will:-

- Improve clinical specific outcomes, e.g. identification and management of respiratory failure, improved nutritional status.
- Improve the quality of Neuromuscular services and patient experience.
- Reduce the number of unscheduled admissions and re-admissions to hospital.
- Reduce hospital length of stay, and thus occupied bed days.

- Improve referral and signposting to appropriate services for patients, including voluntary agencies and community groups.
- Improve transitional care.
- Increase patient choice.
- Reduce inequalities in health between those with neuromuscular condition, both across the region and with other parts of the United Kingdom.

Purpose

Aims

The aims of the services covered by this specification are to

- Provide viable, fully functional, integrated services (i.e. from primary, community, secondary /specialist neuromuscular centres) within each region.
- Provide a holistic and integrated shared care model for individuals with a neuromuscular condition.
- Provide a transitional service that pro-actively and systematically plans care moving from paediatric to adult services.
- Use existing and develop new pathways for standards of care for specific neuromuscular conditions from diagnosis to end of life, ensuring links with primary/community (Tier 1), secondary (Tier 2) and Tertiary tiers (Tier 3) of provision (Tiers 1-3).
- Improve integration of health and social care for patients
- Improve the life expectancy and quality of life of those people living with a neuromuscular condition.
- To centralise the provision of specialised care where necessary and provide care closer to home where possible and appropriate to do so.
- Improve access to education or training for professionals involved in supporting and treating patients with a neuromuscular condition.
- Contribute to training for professionals in recognition of the early symptoms and signs of neuromuscular conditions to reduce diagnostic delay resulting from referral delay.

Service Objectives

The following is a list of service objectives

- Provide improved quality of life and patient experience, for patients and for their carers
- Provide a service that is accessible for individuals and their families, ensuring standardisation of care across the specialist centres.
- Offer the best services within the resources available.
- Provide high quality care for all individuals with smooth transitional care for young adults and effective support for families.
- Patients to receive the majority of their care close to their homes with regular review by local service providers. Specialist centres will act as a coordinator to ensure care plans are in place for local teams to support the patient in the management of their condition.
- Integrated care plans will be used in all cases to ensure lines of communication and whole system management of patients across the tiers of services.

- Specialist services will be available within each region, which are able to support local provision where appropriate.
- End of life care will provide support and care appropriate to the individual and family's wishes.
- Meet national standards and constantly audit the service to document that the best possible outcomes and patient experience are delivered.
- To develop a patient-centred approach to the delivery of services.
- To improve access to services in primary care through referral to more integrated services, particularly for disadvantaged groups and areas.
- To encourage a range of approaches for case-management.
- To encourage greater participation in self-care through appropriate support mechanisms.
- Improve the availability of clinical and non-clinical information.

Scope

Service description

Specialised neuromuscular services will be provided to NMD patients of all ages and via a managed clinical pathway that supports multi-disciplinary and cross organisational working. This will facilitate effective and efficient treatment, care and support to patients, their families and carers.

There are more than 60 different types of muscular dystrophy and related neuromuscular disorders (NMD). Neuromuscular disorders can be genetic or acquired:

Inherited neuromuscular disorders include:

- Muscular dystrophies
- Spinal muscular atrophy (SMA)
- Congenital and syndromal neuropathies
- Inherited neuropathies
- Congenital myopathies
- Metabolic myopathies
- Genetic myasthenic syndromes
- Mitochondrial disorders
- Channelopathies
- Myotonias

Acquired disorders include:

- Myasthenia gravis
- Autoimmune neuropathies
- Inflammatory myopathies.

There are few curative treatment options for most of these diseases. A number, such as Duchenne Muscular Dystrophy, are aggressive and cause progressive muscle wasting and weakness, orthopaedic deformity, cardiac and respiratory compromise, dependency on others for day-to-day care and usually result in premature death. Others (e.g. Hereditary Motor and Sensory Neuropathy) cause

life-long disability without limiting life span.

Some neuromuscular disorders can present in childhood (e.g. Duchenne Muscular Dystrophy) or young adult life. Others can be late onset conditions in adulthood (e.g. inclusion body myositis)

The Specialised Neuromuscular Service will distinguish three key areas;

1. Paediatric
2. Transitional care
3. Adult

Service Delivery

Location(s) of Service Delivery

Each Region will have a directory of services that will highlight:

- Specialist adult and children's neuromuscular services available within the Region
- Specialist neuromuscular respiratory centres that take referrals from the main children's and adult units
- Availability and location of specialist Neuromuscular Rehabilitation Services
- Cardiac centres with an expertise in managing patients with NMD
- Specialist Spinal Surgery Centres

Referral route

Referrals will be accepted from:

- Neurologists
- NMD specialist Nurse
- NMD Advisors
- Long Term Condition Nurses
- Paediatricians
- GP
- Other professionals supporting patients with NM conditions

Pre-diagnosis outpatient care

Children and adults with suspected neuromuscular conditions will be referred by local neurologists or General Practitioners.

Diagnostic screening investigations (e.g. blood Creatine kinase (Ck) levels) are usually undertaken by the local referring team. More specific investigations may be undertaken in consultation with the Neuromuscular Clinic prior to referral. Referral and investigation pathways will be used to avoid delay in specific diagnostic investigations or use of irrelevant or inappropriate tests (see directory).

Specific diagnostic evaluation at the initial neuromuscular clinic will include full neuromuscular examination and/or evaluation by a neuromuscular physiotherapist, followed by investigations which may include metabolic biochemistry, molecular genetic investigations, muscle biopsy, neurophysiology (Electromyography (EMG), nerve conduction studies).

Many patients will need referral to other related services (e.g. respiratory, cardiac) as part of their diagnostic assessment. These referrals should be made directly from the neuromuscular centre. Clinicians will refer for muscle or nerve biopsy in line with agreed clinical referral guidelines. Muscle biopsy analysis may include histochemistry, immunocytochemistry and electron microscopy. The muscle biopsy should be reported by a pathologist with expertise in muscle disease. Nerve biopsy analysis may include histochemistry, immunocytochemistry and electron microscopy. The nerve biopsy should be reported by a pathologist with expertise in nerve disease.

Neuromuscular centres should develop regular multidisciplinary meetings with clinicians, pathologists, clinical chemists and clinical geneticists to discuss the diagnostic pathway and review the results of investigations including muscle biopsy.

Muscle biopsy specimens may need to be sent for review by specialised Neurogenetic Centres (NCG) for the diagnosis of rare disorders. Some molecular genetic investigations are undertaken locally and others by laboratories elsewhere under NCG arrangements.

End of Life Care

End of life care is provided in a range of settings including within the person's own home; within hospices; within hospitals and nursing and residential care. Services should be provided within the most appropriate setting for the individual, and within the models and approaches to best practice as identified in the Strategy for End of Life Care (DH, 2009).

Palliative and End of Life patients will be assessed and identified by their GP/District Nurse (DN), Specialist Nurse, hospital consultant or other appropriate Health Care Professional.

Patients should be referred to their local end of life service and the process in place should trigger standards that are associated with the Palliative Care Gold Standards Framework.

Outpatient review following diagnosis of a neuromuscular condition

The care of all patients should be led from a regional specialist neuromuscular centre with specialist Multi Disciplinary Team (MDT) providing regular local clinics.

The specialist MDT team will comprise of neuromuscular Consultants, neuromuscular physiotherapist, Neuromuscular Nurse, Occupational Therapist, Speech and Language Therapist (SALT), dietician, psychologist and neuromuscular care coordinator. The MDT role will be to assess, diagnose and review patients and their treatment plans. The MDT will also need to facilitate patient access to colleagues including rehabilitation physicians, respiratory physicians, cardiologists, clinical geneticists, orthopaedic surgeons, endocrinologists, gastroenterologists, palliative care clinicians, occupational therapists, and orthotists. These contacts may

not necessarily be at the same clinic appointment but in some circumstances patient centred care will be facilitated if that is possible, so reducing the number of clinics patients need to attend. The MDT will hold meetings to review and discuss new and complex cases prior to a patient's review or new appointment; this may be carried out virtually.

Patients will be reviewed at least every 6 months by the MDT; some patients being treated with steroids or with rapidly progressive neuromuscular conditions will be reviewed every 3 months.

Patients will have integrated care plans in place led and developed by the level 3 centres. The Tier 3 coordinator will establish links with local Tier 2 and Tier 3 services to enable patients to access Tier 1 services to support the continuing management of their condition in between Tier 3 review appointments. MDTs will ensure there are adequate links and communication mechanism between Tiers 3, 2 and 1. For example this might include referral to local or specialised physiotherapy, hydrotherapy, rehabilitation. Through the training delivered by the Tier 3 centres, Tier 1 physiotherapists should have training in both neurological and musculoskeletal physiotherapies.

Patients will have patient held records, updated by those involved in the patient's care. This would enable these to be shared to other professionals who may not be familiar with the patient condition or neuromuscular care for example should a patient present at Emergency Department (ED) . The patient held record would provide contact details of the patients tertiary centre neurology on call services, should a District General Hospital (DGH) or GP clinician require advice and guidance.

Dietary and Nutritional Care

Patients with neuromuscular conditions are frequently prone to nutritional difficulties, including being under- or over-weight.

Patients who have severe weakness may have difficulties chewing or swallowing. This may result in their failing to grow or gain weight in childhood or losing weight in adult life. There is evidence that in patients with neuromuscular conditions general health improves as nutrition improves. Ideal weight is difficult to assess where there is loss of muscle mass. There is some published data giving guidance in some conditions. In all patients, weight should be monitored regularly as part of outpatient review.

Where appropriate other markers of nutrition (e.g. blood iron, vitamin D levels) should be measured. Dietetic advice about calorie dense supplements should be available. In some patients supported enteral feeding via NG tube or gastrostomy may be needed and neuromuscular centres should have referral pathways in place for this. Weight gain is particularly likely to occur as mobility decreases. Patients taking steroids need proactive dietary advice at the initiation of steroid treatment. Neuromuscular clinics need access to means of weighing patients who are not able to stand or sit unsupported. Before a patient is to have major surgery, nutritional

status should be assessed and a plan for potential nutritional support in the recovery phase should be in place.

Respiratory Care

Neuromuscular patients are vulnerable to respiratory complications which may lead to respiratory failure. Neuromuscular clinics need to identify those at risk of respiratory problems and refer for specialist respiratory assessment and monitoring. Identification of those at risk must include an awareness that patients may develop respiratory failure in some conditions whilst they are still ambulant and maintaining good limb strength (e.g. Selenoprotein 1 (SEPN1) myopathy, congenital myopathies including nemaline myopathy, Pompe's disease).

Neuromuscular patients at risk of respiratory failure should be reviewed by the Specialist respiratory failure/Non-invasive Ventilation (NIV) multidisciplinary team every 3 -12 months, depending on the current respiratory status with an open access in the interim to a clinic review within 1-2 weeks. The review will include a respiratory history taking, a focussed examination and physiological assessment for respiratory failure through one or more of Vital Capacity (relaxed or forced), diaphragm muscle strength assessment (Mouth Pressures or SNIP), Peak Cough Flow (PCF), analysis and interpretation of blood gases and acid-base balance. Patients at risk of nocturnal hypoventilation will be offered overnight oximetry or other home respiratory sleep studies to assess for early features of respiratory failure as early treatment with NIV has shown clear benefits in people with neuromuscular respiratory failure and NIV will be actively considered in people with daytime hypercapnia ($p\text{CO}_2 > 6.0$ kPa).

In order to document the progression of a patients progression of respiratory weakness, serial measurement of forced or relaxed Expiratory Vital Capacity (EVC: absolute values and as predicted for height, arm span, or ulna length) should be used. It is important to use the same vital capacity measurement (forced or relaxed) every time so that serial measurements are meaningful.

Once the patient develops clinical signs of nocturnal hypoventilation or EVC drops to 1.25 Litres or <40% predicted value then the serial measurement of overnight oximetry and/or capnography allows the recognition of the development of nocturnal respiratory failure.

The measurement and monitoring of cough flow (PCF) for patients will be used to assess cough strength. Patients will be taught airway clearance techniques as part of their self-management plan to prevent respiratory complications. Lung volume recruitment techniques (e.g. breath stacking bags or via NIV) and techniques to augment cough (manually assisted cough) should be taught when the PCF is <270 l/min/FVC<40% predicted in non-ambulant patients and should be introduced before the patient is less than 160 l/min (ATS 2004)

If a patient's EVC begins to drop they may be susceptible to respiratory infections and should be offered influenza, pertussis and pneumococcal vaccination and when coughing is ineffective, antibiotics should be provided promptly, in selected cases as a supply of a "rescue" course.

If a patient's coughing continues to be ineffective and PCF is around 160l/min/FVC <40%, cough assist machines should be introduced to increase PCF and thereby assist secretion clearance (Lancet 2009). This should also be taught and used in all neuromuscular patients with PCF <270l/min who are undergoing anaesthetic/sedation, to prevent post-operative respiratory complications (Birnkrant 2007).

Symptomatic nocturnal hypoventilation is an indication for the initiation of elective non-invasive nocturnal Non-invasive ventilation (NIV) and symptoms of this should be checked at every visit as specified in clause 3.4.2 above. NIV should also be considered if Respiratory Sleep studies and other physiological assessments warrant. As nocturnal hypoventilation can start without daytime symptoms and as individuals' thresholds of tolerating symptoms vary widely, a discussion about the advantages and disadvantages of nocturnal NIV should commence at the point at which physiological abnormalities are identified (daytime hypercapnia $pCO_2 > 6.0$ kPa) and not wait for symptoms to develop.

The Specialist respiratory failure/Non-invasive Ventilation (NIV) multidisciplinary team should provide structured education to the patient and carer in use of ventilation equipment.

Cardiac care

Neuromuscular patients are vulnerable to cardiac involvement, which may be as manifest as cardiomyopathy or as rhythm disturbance. The risk of development of such cardiac complications is extremely high in some conditions (e.g. Duchenne MD), possible in others (e.g. myotonic dystrophy DM1) and not increased in some (e.g. Charcot-Marie-Tooth neuropathy 1A (CMT1A)).

All patients with a neuromuscular condition known to associated with a risk of cardiac complications should be referred for a cardiac assessment. This will include women who are carriers of a Duchenne muscular dystrophy gene mutation. Cardiac review should be considered in all patients who have an uncategorised myopathy where the risk of cardiac involvement is unknown but possible.

Neuromuscular patients with cardiac involvement should be reviewed at regular intervals by a consultant Cardiologist with specific expertise in the cardiac complications of neuromuscular disease. These conditions are rare and expertise will be maintained and developed if these patients are seen in a specialised cardiology clinic. There is a need for follow up cardiology examinations at intervals in at risk patients even if the initial investigations are normal as cardiac complications can evolve over time.

Orthopaedic and spinal care

Patients with muscle weakness are vulnerable to develop spinal deformity, especially scoliosis, particularly in childhood or adolescence. The risk of scoliosis is high in Duchenne muscular dystrophy, Spinal muscular atrophy, congenital muscular dystrophy and some forms of congenital myopathy. Patients should be assessed for the development of a scoliosis when attending the neuromuscular clinic. Parents of

children who are at risk of scoliosis development should be made aware that in some children this can develop and progress rapidly over a period of months and this may be a reason for review earlier than planned. If a scoliosis or other spinal deformity (e.g. kyphosis) is present, patients should be referred to a specialist spinal team for an outpatient assessment and consideration of suitability for surgery. Assessment of suitability for surgery will also depend on information from respiratory and cardiac assessments.

Patients with neuromuscular conditions frequently develop orthopaedic complications which contribute to disability in children and adults. These may include progressive foot deformity, joint contractures and joint instability or subluxation. Patients should be referred for an orthopaedic assessment. This may be undertaken by local orthopaedic services in many instances but some patients will need specialist orthopaedic input from clinicians with expertise in specific conditions.

Wheel Chair Services

For the provision of wheelchairs, a patient's Physiotherapist or Occupational Therapist should make the decision with referrals falling in line with local clinical Commissioning Group (CCG) referral guidelines

Orthotic provision

Orthotic advice and support will be arranged as and when this becomes necessary. Care coordinators will facilitate referrals to local or specialised services as appropriate to the needs of the patient.

Psychological Support

Care coordinators will have a key role in supporting patients and carers with both the emotional and practical problems that arise for those living with neuromuscular conditions. Social (information, advocacy and advice) and psychological support are needed particularly at times of changing needs and crises, for example, moving/adapting house, loss of ambulation, surgery, the development of cardiac & respiratory problems, starting university/employment, and end of life.

Neuromuscular clinics should have access to psychological support for children and adults and develop expertise with a practitioner experienced in the specific challenges for neuromuscular patients. This should be available for patients and families for instance at times of crises as above and for children with behavioural issues, which may be a recognised feature of some conditions (e.g. Duchenne muscular dystrophy), arise as a complication of treatment (e.g. steroid therapy), or be precipitated by the challenges of adjusting to progressive disability. The neuromuscular service will offer links to independent patient support groups, and support their own patient groups. Such groups are a vital source of peer support, advice and information for patients. They may also be able to act in an advocacy role, either at group or individual level, and as agents for change and service improvement.

Shared care plans with local services should consider family referral to local psychology services to support them as they come to terms with the diagnosis.

Access to Rehabilitation and Specialist Equipment

Referral to a local rehabilitation service should be considered for patients with neuromuscular disorders. The interval between assessments depends on the specific diagnosis, the progression of the disease and the patient's functional ability. The aim of the assessments is to plan for interventions for the patient to optimize their physical, social, vocational and intellectual abilities. Referral to specialised regional rehabilitation (where available) should be made as appropriate.

Patients should be referred for enabling equipment such as wheelchairs adaptations and environmental controls in line with CCG or specialist rehabilitation referral criteria. The use of integrated care plans should ensure appropriate referrals are made and all professionals involved in the patient's care are aware of the equipment needs. There should be clear guidelines defining the local provision of suitable equipment to support patients 'independence, functionality and to reduce the risk of complications. For the provision of wheelchairs, a patient's Physiotherapist or Occupational Therapist should make the decision with referrals falling in line with local PCT referral guidelines.

Much of the equipment and rehabilitation care can be provided locally but the regional posture and mobility services can be consulted when local services do not have the expertise or require further specialist opinion regarding suitable wheelchair provision for a particular patient.

Transitional care

The tier 3 neuromuscular clinic Care coordinator will support the transition to adult services. The Care coordinator will develop a transition plan for young people with a neuromuscular condition moving from paediatric to adult services. Once a patient reaches the age of 13 then discussions and planning for the patient's future should begin.

Other

Evidence Base

- Historically a number of regions have seen the development of specialist centres of excellence based on university research departments or existing specialist centres e.g. Newcastle, London, Oxford and Oswestry.
- Over the last few years' diagnosis and management of neuromuscular conditions have improved, increasing life-span so that patients who had previously survived only into childhood are now becoming adults.
- The introduction of standardised care that includes multidisciplinary team review with respiratory monitoring and support, active cardiac management, orthopaedic interventions and the use of steroids in Duchenne muscular dystrophy, have all contributed to this improvement in survival rates. However, increasingly complex care packages and comprehensive local support for

patients and their families requires specialist support.

- The Treat NMD consortium – a European Union Network of Excellence for the development of translational research in rare neuromuscular diseases - has published and disseminated international consensus on care and management for patients with Duchenne muscular dystrophy, Spinal Muscular Atrophy and Congenital muscular dystrophies. Treat NMD is an international initiative bringing together some of the world's leading neuromuscular specialists in a pan-European 'network of excellence' aimed at improving treatment and finding cures for patients with neuromuscular disorders.
- International Guidelines recommend cardiac care in patients with Duchenne Muscular Dystrophy (DMD) or Becker Muscular Dystrophy (BMD), which includes details of the cardiac assessment and clinical management by a cardiac specialist with an interest in the management of cardiac dysfunction and/or neuromuscular disorders.
- International standards of care for Duchenne Muscular dystrophy, including diagnosis, pharmacological and psychosocial management and the implementation of multidisciplinary care were published in 2010 (Part 1 Lancet Neurol. 2010 Jan; 9(1):77-93. Part 2: Lancet Neurol. 2010 Feb; 9(2):177-189)
- Standards of care for Spinal Muscular Atrophy were published in 2007 (J Child Neurol. 2007 Aug; 22(8):1027-49)
- An international consensus statement on Standard of Care for Congenital Muscular Dystrophies was published in 2010 (J Child Neurol 201,25;15591581)

Exclusion Criteria

Where patients have been assessed and their needs are not specialist in nature and can be cared for locally.

Discharge Criteria & Planning

As this is a service for people with long term, deteriorating conditions, discharge is rare. If patients feel they are sufficiently supported locally and are stable, they will be given open appointments for them to be reviewed as needed.