



PAEDIATRICS

A NATIONAL MODEL OF CARE FOR PAEDIATRIC HEALTHCARE SERVICES IN IRELAND

CHAPTER 34: PAEDIATRIC NEUROLOGY AND EPILEPSY



Féidhmeannacht na Seirbhíse Sláinte
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**ROYAL
COLLEGE OF
PHYSICIANS
OF IRELAND**

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34.0 OVERVIEW

This model of care for paediatric neurology builds on the work outlined in the Laffoy report Neurology Services in the Strategic Review of Neurology and Neurophysiology (2007) and takes account of the Neurology Needs Assessment – Paediatric Neurology and Neurophysiology (King et al., 2007) which was approved by Warlow and van Gijn in 2009 (Warlow and van Gijn, 2009). The key concept proposed in this document, i.e. that of a managed clinical network for neurology, was initially introduced in the Laffoy report. It is proposed that, within the clinical network, there will be a national quaternary/tertiary service specialising in the management of complex conditions, including paediatric neurointensive care at the new children's hospital in Dublin; a large regional neurology service in Cork; secondary neurology services in Limerick and Galway; and local neurology services in all other paediatric units working together towards best patient outcomes. The realisation of the managed clinical network for neurology, as outlined in this model of care, is integral to achieving the vision of a world-class paediatric neurology service in Ireland.

This document should be read in conjunction with other relevant models of care developed through the Health Service Executive (HSE) National Clinical Programmes and Strategy Division, in particular, the models of care for epilepsy (HSE, 2015), neurology (HSE and RCPI, 2015) and the paediatric neurodisability chapter in this national model of care for paediatrics and neonatology. Together, these models of care cover the continuum of care for patients with neurological conditions from acute management, diagnosis, hospital-based services (including therapy), specialist rehabilitation and continuing care in the community led by specialist neurorehabilitation teams.

34.1 INTRODUCTION

Paediatric neurology is a quaternary/tertiary specialty which must be geographically distributed so as to allow all children equal access to care. The service model must enable smooth delivery of a specific care pathway for each individual child or young person with neurological disease. The provision of care is designed and led by the consultant paediatric neurologist. The recommended number of paediatric neurologists in developed countries ranges from 1/100,000 to 3/100,000 children aged under 16 years. In 2014, (BPNA, 2014) there were approximately 115 whole-time equivalent (WTE) consultant paediatric neurologists in the National Health Service (NHS) in the United Kingdom (UK), compared with 61 in 2002. There were 13 WTEs in Scotland, whose population and demographics are similar to Ireland's, but which has more uniform community paediatric services.

The aim of the paediatric neurology service is to ensure that children and young people with serious neurological conditions achieve the best quality of life, through the provision of excellent diagnosis, investigation, intervention, management and information, as close to home as possible.

The objectives of the service are:

- Provision of accurate diagnosis of children with neurological disorders including the expert management of life-threatening and potentially treatable disorders, the avoidance of disability through delayed or inappropriate treatment and the avoidance of further affected cases through recognition of genetic disorders.
- Maintenance of appropriate guidelines for management of common conditions such as epilepsy, status epilepticus, neonatal seizures, headache, developmental delay.

- Provision of specific programmes of case-based management of complex neurological disorders such as epilepsy surgery for intractable epilepsy or new genetic therapies in neuromuscular disease.
- Provision of a training programme in paediatric neurology with involvement in leading-edge research, and ongoing education and training for all members of the paediatric neurology team.
- Establishment of pathways of care in liaison with other subspecialties including neurosurgery, rehabilitation, genetics and child development, and ensuring a smooth transition of care for the adolescent with a chronic neurological disorder to the adult service.
- Introduction of new initiatives such as the electronic patient record for children with epilepsy, the maintenance of national databases for children with neurological disorders, and participation in international audit such as the 'Epilepsy12' national audit in the UK.

International Observations and Recommendations

Paediatric neurology is a broad specialty involving the diagnosis and treatment of a wide range of disorders, as listed in Appendix 1. It has been estimated that children with neurological disorders account for up to one-third of patients referred to a district general hospital, and that a quarter of all paediatric admissions to hospital would benefit from a paediatric neurology opinion. Paediatric hospital discharges coded with a primary non-surgical neurologic ICD-9 code may account for up to 15% of paediatric discharges from an academic medical centre (Nash et al., 2013).

However, it is estimated that only one-third of children and young people with neurological disease are referred to specialist centres for diagnosis, evaluation and treatment. Furthermore, the lifetime care of children and young people with complex, usually neurologically-based, disability (whether congenital or acquired as a result of illness or trauma) is largely managed by community-based or hospital-based local paediatricians working in multidisciplinary teams (MDTs), with access to more specialised care on a consultation basis. Increased expectations engendered by advanced information technology, improved standards of living and education, and ease of international travel mean that families whose child has a significant neurological symptom expect to gain speedy access to a child neurologist for consultation and diagnosis. The impact on a family of a diagnosis of a chronic neurological disorder in a child is enormous and a high rate of marital breakdown is associated with such diagnoses.

Each service must provide a service to a population of sufficient size, in order to maintain the clinical, practical and academic viability of the unit. The London Commissioning Review noted that most world-leading providers of specialised services for children served a population of at least 3.5 million. That review concluded that, in the case of London, three or more networks would be unlikely to provide the necessary critical mass and interdependencies between services; it therefore recommended two networks, each of which would include a lead hospital and a number of other hospitals that delivered agreed specialised children's services across their network. Based on this recommendation, Ireland should have one managed clinical network in paediatric neurology coordinated through the new children's hospital in Dublin.

The majority of paediatric neurology services are recognised as specialised (otherwise known as quaternary/tertiary level) and, consequently, are based in neurosciences centres which have the necessary infrastructure in terms of diagnostic services and other specialties. The service aims to provide all treatments from the point of referral. Ideally, referrals are made by the doctor to a consultant paediatric neurologist for acute or chronic referrals, and second neurology opinions should be requested by the attending paediatric neurologist. The majority of patients cared for by paediatric neurology services are referred from secondary-level paediatric services. Others

are referred directly from primary care, within agreed protocols, or from other paediatric specialties, including paediatric neurosurgery, and orthopaedics. Paediatric neurology should provide access to subspecialty care through:

- (a) ready contact with secondary-level general paediatric services where uncomplicated epilepsy, migraine, and neurodisability are usually managed locally
- (b) outreach, outpatient services based in secondary care centres around the geographical region served.

This allows specialist services to be provided as near to patients as is reasonably feasible. Patients seen in outreach clinics are usually under the joint care of a local paediatrician with or without subspecialist interest in paediatric neurology.

Most paediatric neurology consultation is delivered in an outpatient setting in a hub-and-spoke model. Out-of-hours advice, including access to direct clinical neurological assessment, is required for the care of those children and young people admitted to the care of neurologists as inpatients, and those with neurological problems as part of complex illness who may be cared for in paediatric intensive care units (PICUs) and/or under the care of paediatric specialties other than neurology. Consultation to other inpatient units in the clinical network is provided by telephone.

Paediatric neurology services are provided in both outpatient and inpatient settings. Outpatient services are provided from a range of outpatient clinics, which should include general paediatric neurology clinics and subspecialist clinics, such as epilepsy, neuromuscular, neurocutaneous, movement disorders, demyelination, neonatal neurology, narcolepsy and neuro-oncology clinics. Many such clinics are multidisciplinary, involving other medical specialists, therapists and other allied professions. Children requiring diagnostic investigations often have these undertaken as day case admissions.

Investigations and Diagnosis

Diagnostic facilities necessary for the care and management of children with neurological disorders include:

- Haematological, biochemical and microbiological investigations
Paediatric neurological services require access to high-quality haematological, biochemical and microbiological investigations to detect and monitor disturbances in body systems outside the brain and to investigate infectious diseases (viral, bacterial, protozoal, fungal and prion), both of the nervous system and outside the nervous system.
- Imaging
Paediatric neurology services require ready access to conventional x-ray imaging of skull, spine and other bones (including skeletal surveys for suspected non-accidental injury); digital subtraction angiography; computerised tomography (CT) brain (including CT angiography); magnetic resonance imaging (MRI) brain (including magnetic resonance (MR) angiography, MR spectroscopy and functional MRI) and MRI spine; neonatal brain ultrasound examination and Doppler ultrasound vascular imaging. In addition, paediatric neurology may require access to positron emission tomography (PET) and single photon emission computed tomography (SPECT). Such specialised imaging may not be available on site and may need to be accessed at a supra-regional centre.

- **Neurophysiology**
Electroencephalography (EEG) (including standard and sleep recordings; ambulatory EEG and EEG video telemetry; evoked potentials (visual, brain stem/auditory; somatosensory and transcranial motor – which form the basis of neuro-intraoperative monitoring (NIOM) for spinal surgeries); nerve conduction studies; electromyography (EMG) and electroretinography (ERG).
- **Neurometabolic investigations**
The ability to investigate potential neurometabolic disorders is essential to paediatric neurology. These investigations are mainly done on blood, urine and cerebrospinal fluid (CSF). Less commonly, they are performed on samples derived from biopsies taken from skin, liver, bone marrow and brain. Many neurometabolic investigations will be processed on site but sent to specialist supra-regional, national or international laboratories.
- **Neuroendocrine investigations**
The ability to investigate potential neuroendocrinological disorders is essential to paediatric neurology. These investigations are mainly done on blood samples and occasionally on CSF. Some neuroendocrine investigations will be processed on site but sent to specialist supra-regional or international laboratories.
- **Neuroimmunological investigations**
The ability to investigate potential neuroimmunological disorders is essential to paediatric neurology. These investigations are done on blood and CSF samples. Some neuroimmunological investigations will be processed on site but most are sent to specialist supra-regional or international laboratories.
- **Neuro-ophthalmological investigations**
Ready access to specialist paediatric ophthalmological services that can provide direct and indirect fundoscopy, optometry, ocular ultrasound and visual field testing is essential for paediatric neurology services.
- **Neuropathological investigations**
Ready access to a neuropathology laboratory that can process and analyse CSF cystospin specimens, and biopsy material taken from brain, muscle, nerves and other body organ and tissues is necessary for paediatric neurology. In addition, access to supra-regional, national and international neuropathology laboratories is sometimes required, particularly in the investigation of children with neuro-oncological, neuromuscular and neurodegenerative disorders.
- **Genetic investigations**
Ready access to clinical and laboratory genetic services that can provide comprehensive clinical, chromosomal and molecular DNA diagnostic genetic services is essential for paediatric neurology services.
- **Psychometric testing**
Paediatric neurology services require access to psychologists and neuropsychologists who can undertake psychometric evaluations in children of all ages and in children with various impairments (including visual and hearing impairments).

- **Cardiological investigations**
Ready access to 12-lead echocardiogram (ECG) is required for the investigation of many children with paroxysmal events. Access to tilt testing is required for the investigation of selected children with paroxysmal events. Cardiological assessment, including echocardiography (and trans-oesophageal echocardiography), may be required in the assessment of children presenting with strokes and brain abscesses and in children with neuromuscular diseases.
- **Respiratory investigations**
Access to lung function tests and overnight saturation-monitoring sleep studies is required for the investigation and management of some children with neurological conditions, especially neuromuscular diseases.
- **Gastrointestinal investigations**
Oesophageal pH monitoring and videofluoroscopy.
- **Speech and language assessment in regression, aphasia, feeding and swallowing difficulties** is required for the management of many children with neurological disorders.
- **Audiological assessment**

Investigations are carried out in a variety of settings, including outpatient departments, day case units and inpatient units. Paediatric emergency neurology services need to be able to access many investigations facilities out of normal working hours. These include haematological, biochemical and microbiological services, conventional x-ray services, brain CT and MRI and, increasingly, EEG. Access to out-of-hours neuropathological services, including processing of samples to be analysed later, is occasionally required. Certain investigations, particularly neuroimaging, may require general anaesthesia in some patients. In order to provide an acute paediatric neurology service, 24-hour access to general anaesthetic services is necessary.

Care Settings

Inpatient Care

Paediatric neurology inpatients are located in a variety of settings, including designated paediatric neurology beds (often in a joint neuroscience ward shared with paediatric neurosurgery), paediatric intensive care beds and on neonatal units. Inpatients may be under the joint care of other consultants, especially general paediatricians, paediatric intensivists, cardiac intensivists, neonatologists, metabolic paediatricians and paediatric neurosurgeons. Most inpatients will have acute neurological disorders or acute medical problems caused by an underlying neurological disorder. Hospital admission will have been necessary in order to enable diagnostic tests only reasonably available in an inpatient setting to be undertaken, or to allow inpatient-based therapeutic interventions to be undertaken. Many such patients will need to be nursed in an intensive care setting or in a high-dependency unit. Inpatients with disorders principally involving other body organs and systems often have associated neurological problems or complications. Paediatric neurology services provide consultation services to such children.

Ongoing Care

Children who recover from acute or long-term neurological conditions may be discharged back to the care of their general practitioner (GP). Children with ongoing neurological problems not requiring ongoing specialist neurological services may be discharged to general or community paediatric services or neurodisability services. This includes children who require, or have required, neurorehabilitation.

Neurological Conditions

The range of conditions attending the paediatric neurology service is outlined in Appendix 1, with comments on the role of the neurologist and secondary care paediatrician, where appropriate. The conditions which should be excluded from paediatric neurology services are listed in Appendix 2.

Documents are provided as an appendix or as a key reference on the following:

- Requesting neurophysiology (EEG, EMG, evoked potentials) (Appendix 3)
- Guideline for treatment of Status epilepticus >3 months (Appendix 4)
- Guideline for treatment of Status epilepticus <3 months (Appendix 5)
- Out-of-hospital rescue medication (Appendix 6)
- Neuromuscular model of care (Appendix 7)
- Developmental delay (O'Byrne et al., 2015)
- Narcolepsy (Allen et al.) (Appendix 8)
- Headache (Appendix 9)
- Neurofibromatosis (Scottish Intercollegiate Guidelines Network, 2005)
- Status dystonicus (Allen et al., 2014)
- Ketogenic diet pathway (Appendix 10)
- Paediatric population/admissions Census 2011 (Appendix 11)

Epilepsy

The prevalence of epilepsy in childhood makes it suitable for development of a paediatric neurology model of care which has been described in detail in the epilepsy model of care (HSE, 2015). This model will continue to develop and modify care pathways based on best practice and international guidelines such as those of the National Institute for Health and Care Excellence (NICE) (NICE, 2004 and NICE, 2012) and the Scottish Intercollegiate Guidelines Network (SIGN) (Scottish Intercollegiate Guidelines Network, 2005).

The key features are outlined here:

- Paediatric epilepsy is the most common serious neurological disorder with an overall childhood incidence of 50-70 cases per 100,000 per year and a childhood prevalence of 5-10 cases per 1,000 population. This affects about 0.7% of all children.
- Societal costs are considerable as individuals with medically intractable seizures make up one-third of the epilepsy population.
- The vision for the programme is to provide the best value care for all children and adolescents with epilepsy in the right place, at the right time, sharing the best available information. Central to this integration is that it will provide timely access, intelligent support and outreach clinics for general paediatricians nationally.
- A Cochrane review of models of epilepsy care found strong evidence for positive outcomes by the involvement of epilepsy nurse specialists and self-management strategies alone. Outcomes measured included morbidity, mortality, seizure freedom, quality of life and knowledge of epilepsy and its treatment.

Management of epilepsy in children may be difficult for the following reasons:

- Diagnostic difficulties may arise (other disorders masquerading as epilepsy).
- Investigations often require sedation/anaesthesia and may be normal early on.
- Many epilepsies are age-related syndromes requiring specialised treatment.
- Children with epilepsy require extensive multidisciplinary (neuropsychology, psychology, occupational therapy, school support) input throughout their childhood years.
- Tests for surgical evaluation may be invasive

Managed clinical networks are very effective in bringing together key professionals, establishing well-defined, integrated care pathways and clinical guidelines for each step of the pathway.

The resources required locally include:

- primary care physicians
- local paediatrician
- clinical nurse specialist (CNS) in the region with expertise and responsibility for epilepsy
- local access to psychology, speech and language therapy, occupational therapy, physiotherapy
- access to EEG and neuroimaging – the network should have guidelines for requesting these studies
- information for families on investigations, epilepsy, syndrome diagnosis, and treatment – including surgery
- neuropsychology and child psychiatry as needed

The potential impact of a managed clinical network for children with suspected epilepsy is immense:

- more direct access to specialist advice, diagnosis and treatment
- lower rates of misdiagnosis
- fewer unwanted side effects from drugs
- increased cure rate through selection for epilepsy surgery
- earlier intervention by educational and social services
- elimination of wrong diagnosis of epilepsy for some children

Working in tandem with paediatric neurology services in Dublin, Cork, Limerick and Galway the network will provide rapid access service for community and emergency department (ED) referrals, telephone, email and web-based advice. The delivery of care will have an emphasis on co-managing with local paediatricians. The paediatric services will also benefit from transition clinics, to introduce adolescent patients with epilepsy to the staff on the adult service. Through the use of an integrated care pathway, administered through the ED, front-line paediatricians will use an intelligent, evidence-based algorithm to help with management and disposition of patients who come to hospital with first seizures or breakthrough events. International evidence suggests that too many patients are being admitted when, in many instances, they could safely receive care and advice and rapid follow-up as an outpatient. Front-line paediatricians delivering care at the ED interface will be provided with an integrated seizure care pathway that will reduce admissions and length of stay, while improving patient safety by eliminating treatment variability (see model of care for epilepsy).

The 15% or so of patients with highly complex epilepsy who need to be assessed for possible epilepsy surgery will be dealt with by increasing the number of paediatric pre-surgical evaluation beds in Dublin to four.

Access

- The Programme aims to improve support to general paediatricians in providing paediatric epilepsy care.
- This support will be based on:
 - a. guidelines for management of common epilepsy syndromes (see epilepsy model of care and NICE Guidelines).
 - b. improved web-based access to epilepsy centres in Dublin and Cork
 - c. improved access to regional advanced nurse practitioners (ANPs) in epilepsy.
- Managed clinical networks, anchored by general paediatricians in paediatric units around Ireland, will be developed.

- These clinical networks would be centred around two epilepsy centres in Dublin and Cork, respectively, and would function on a hub-and-spoke model.
- Following the appointment of additional paediatric neurologists, a programme of outreach clinics to peripheral units – run by a visiting paediatric neurologist, an ANP and an EEG clinical measurement scientist – would be developed.

Quality

- improvement in the accuracy of diagnosis in childhood epilepsy by hosting further paediatric epilepsy training courses and through greater access to paediatric neurology
- improvement in seizure freedom through more accurate diagnosis and more rational drug use
- reduction in bed days in paediatric epilepsy care

34.2 CURRENT SERVICE PROVISION

The paediatric neurology service began in the 1960s when Professor Niall V. O'Donohoe was appointed to Our Lady's Children's Hospital, Crumlin (Crumlin). A second centre was established in Temple Street Children's University Hospital (Temple Street) in 1987, followed by the unit in Cork University Hospital in 2004.

Current Clinical Paediatric Neurology Consultant Manpower in Ireland

The number of paediatric neurologists is currently 6 WTEs with a further post to be filled in Dublin in late 2015.

- In Dublin, the 4 neurologists provide 24/7 on-call service (1 in 2 rota over 2 sites; 1:1 when colleague on leave) including neurointensive care and neonatal neurology. This Dublin service is the only 24/7 on-call service for paediatric neurology in Ireland.
- In Cork there are 2 WTE consultant paediatric neurologists.
- In Limerick there is a temporary sessional service in paediatric EEG and epilepsy from a consultant paediatrician with subspecialist interest in paediatric neurology.
- In Galway, the recently appointed professor of paediatrics has subspecialist interest in paediatric neurology.

Current Consultant Clinical Paediatric Neurophysiology Manpower in Ireland:

- In Dublin there are two WTE paediatric neurophysiologists (one each in Crumlin and Temple Street).
- In Cork, neonatal and paediatric EEG are reported by an adult clinical neurophysiologist with paediatric expertise.

Referral Pathways

While there is some variation in referral pathway across sites, most referrals for both paediatric neurology and neurophysiology evaluation come from GPs, general paediatricians, child and adolescent mental health services (CAMHS), or community area medical officers. The referrals are triaged by the consultant and prioritised according to degree of urgency. Where indicated, e.g. new onset seizures, contact is made with family by a CNS/ANP. Direct consultant-to-consultant contact is usual for those urgent referrals/transfers from hospitals outside Dublin. Inappropriate referrals are returned/redirected with contact from the team as appropriate.

Main Deficits in Paediatric Neurology Services in Ireland

There is enormous variation in resources, support services and neurophysiology support services provided across the sites. This is summarised below:

(a) Inadequate consultant paediatric neurology posts

- There are unacceptable waiting times for non-urgent outpatient first attendances to neurology clinics, ranging from 6-24 months.
- Outpatient clinics are too large with patients not getting recommended allocation time of 15-20 minutes (return visit) or 40-60 minutes (first visit).
- There are no outreach clinics, with families from sparsely populated regions (Central Statistics Office, 2011) making up to 12-hour return journeys for review at an outpatient clinic (e.g. Belmullet to Dublin).
- There are no managed clinical networks.
- There is no academic post in paediatric neurology (unique in Western Europe).
- There is no higher specialist training programme in paediatric neurology in Ireland.
- There are no national registers or databases in paediatric neurology, vital for epidemiology, planning services, participating in research and therapeutic trials as treatments are developed.
- There is a major difficulty recruiting consultant medical staff to the service, in part because of perceived workload, a problem recognised worldwide (Ferriero and Hauser, 2010; Ridel and Gilbert, 2010), but only recently an issue in Ireland.
- There is little audit or research.
- There are no national distance learning courses for medical or nursing staff.

(b) Inadequate support

- There is only one WTE paediatric neuropsychologist in Ireland (0.5 WTE Temple Street and 0.5 WTE Crumlin).
- There are unacceptable waiting times for assessment and treatment in services from speech and language therapy (SLT), occupational therapy (OT), physiotherapy and psychology at community, secondary and tertiary levels. Community services need to be developed appropriately, so that children receive the required services as close to home as possible, which will reduce the burden on secondary and tertiary services in some instances.
- Dietetic services are totally inadequate across all units for number of patients who might benefit from a) ketogenic diet in epilepsy or b) feeding evaluation in children with complex disorders.
- Community child health and neurodisability services are very unevenly distributed throughout Ireland.
- CAMHS are totally inadequate across all units, with one service experiencing the collapse of a neuropsychiatry liaison service in 2013.
- Cork inpatient child psychiatry service is provided by the community CAMHS team only to patients residing in Cork city; thus patients who reside outside Cork city do not have access to inpatient assessment.
- Activity levels from hospital inpatient enquiry (HIPE) and outpatients do not capture all activity, especially inpatient consultation service, which is a major part of the paediatric neurology service.
- General paediatrics is limited, especially in the Dublin hospitals, leading to inappropriate review attendances at neurology clinics.

(c) Inadequate neurophysiology support

There is a requirement for a third clinical neurophysiology post within the new children's hospital structure to service paediatric neuro-intraoperative spinal monitoring. Approximately 200 cases of paediatric spinal deformity require surgery annually. At present, there is a lack of specialist expertise in the provision of spinal monitoring to children and the service is provided by private contract with a UK consultant. The third post would have responsibility for the development and delivery of a comprehensive service in paediatric intraoperative spinal monitoring.

34.3 PROPOSED MODEL OF CARE

Clinical Paediatric Neurology

An integrated, nationwide, equitable paediatric neurology service with centralised expertise leading to centres of excellence and peer review, and avoiding isolation of specialists, should have 1-2 consultant paediatric neurologists per 100,000 children plus all the administrative, nursing and health and social care professional support as outlined below.

The service should be delivered by a managed clinical network supported by the National Neurology Steering Group with elements such as distance learning programmes and paediatric epilepsy training courses, in addition to outreach clinics. Each satellite paediatric centre should have at least one local paediatrician and one nurse with responsibility for epilepsy ± disability (as many children with complex epilepsy will have disability).

Based on the 2011 Census (Central Statistics Office, 2011) childhood population distribution (see Appendix 11), there should be:

- a single tertiary/quaternary centre at the new children's hospital in Dublin
- a large regional centre at Cork University Hospital
- smaller regional services based in Limerick and Galway
- Single tertiary/quaternary centre at the new children's hospital in Dublin. This centre would have 8-10 consultant paediatric neurologists (including an academic post), all with general training (particularly epilepsy) but with subspecialisation such as:
 - epilepsy (including pre-surgical evaluation)
 - neuromuscular disorders
 - movement disorders
 - cerebral vascular disease
 - neurometabolic disease
 - neuroinflammatory disorders
 - neonatal neurology
 - neurocritical care
 - neurogenetics
 - neurocutaneous disorders
 - academic: any of the above areas of subspecialisation

This national paediatric neurology service would provide a national service in complex epilepsy care, epilepsy surgery evaluation, neuromuscular disease, movement disorders, multiple sclerosis, narcolepsy in addition to neurointensive care for complex acute encephalopathies, myelopathies, vasculopathies, neuromuscular disorders,

among others. The service would provide second opinions on undiagnosed patients attending neurology services outside Dublin through disease-specific clinics. The Dublin service will serve a population of approximately 659,462 children, in addition to its national service.

The Dublin service would establish a network with the large regional centre in Cork, the regional centres in Limerick and Galway, and develop outreach clinics at other secondary care units at Sligo/Letterkenny, Cavan/Drogheda, Portlaoise/Mullingar, and Kilkenny/Wexford as part of a managed clinical network working in parallel with the telelink services being developed in EEG and neuroimaging. Referrals to this service would come from:

1. General paediatricians, or paediatricians with a subspecialist interest in paediatric neurology, or paediatric neurologists.
 2. GPs in the Greater Dublin area with reference to the guidelines in Appendices 1 and 2.
 3. Second opinions should only be requested by the attending neurologist to ensure appropriateness of diagnosis and avoid duplication of investigations.
- Large regional centre at Cork University Hospital. This should be tri-located with both an adult hospital and a tertiary neonatal unit and would serve a childhood population of 193,226 (Cork, Kerry, South Tipperary, Waterford). This centre would have three consultant paediatric neurologists, all with general neurology training but also with subspecialisation such as epilepsy and neuromuscular disorders. This service would link in with the paediatric units in Munster (Tralee, Clonmel, Waterford) through outreach clinics and the managed clinical network at the new children's hospital in Dublin.
 - Smaller regional services based in Limerick and Galway. Each centre would have one WTE consultant. The service in Limerick would serve a childhood population of 85,535 (Limerick, Clare, North Tipperary). The service in Galway would serve a childhood population of 98,744 (Galway, Mayo, Roscommon). The recent appointment of a paediatrician with a subspecialty interest in paediatric neurology to the chair of paediatrics in UHG/NUIG will facilitate the development of a managed clinical network linked to the West.

There may also be a requirement for a paediatrician with a specialist interest in paediatric neurology in Sligo/Donegal/Leitrim (population served 60,536). However, as the population served is small, it could be argued that the low volume of patients would lead to difficulty in maintaining skills and expertise, compounded by a degree of isolation from other professionals and developments in this field. The need for this post will be ascertained during the initial phase of establishing the network.

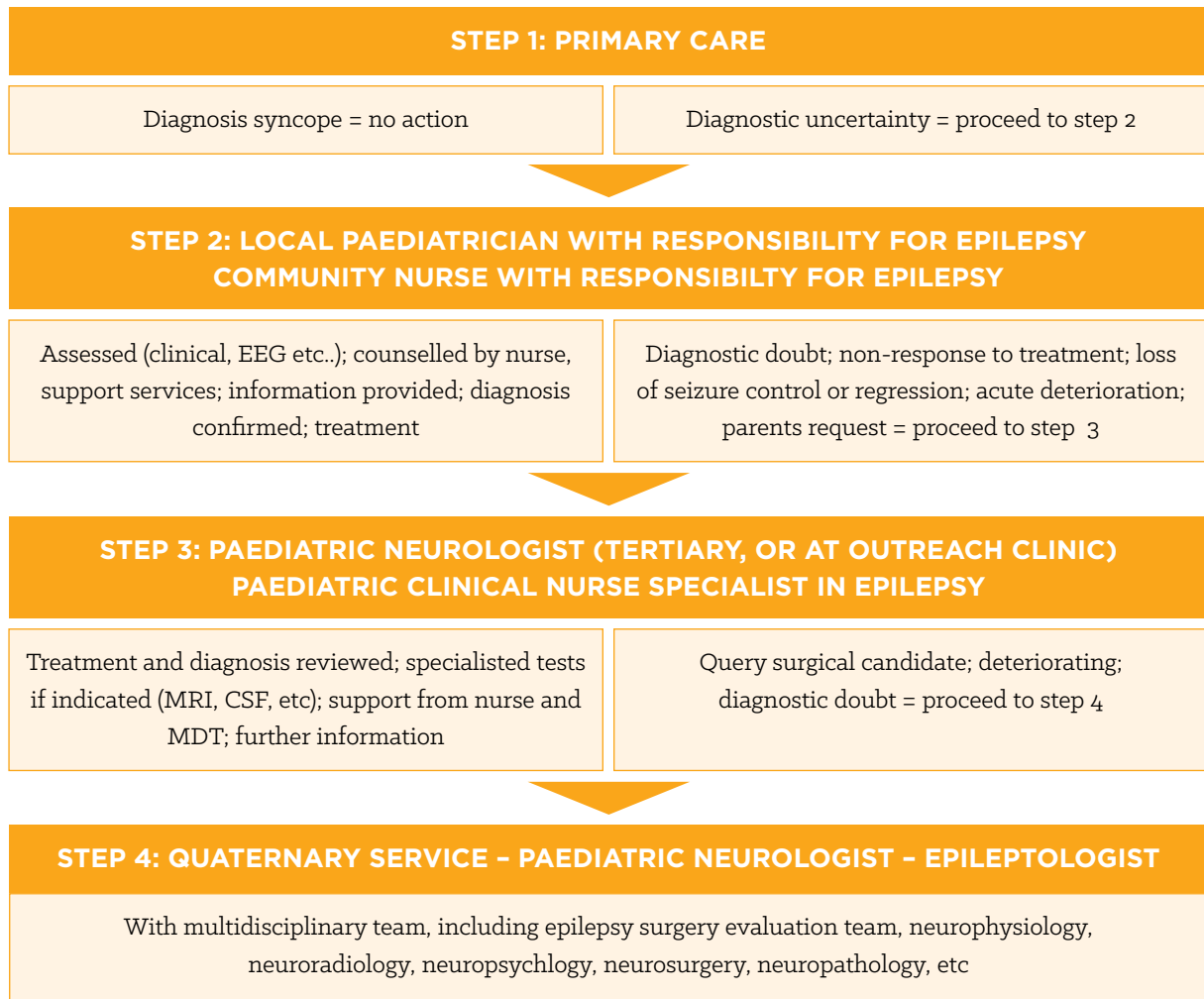
These centres, all with existing adult neurology services, would act as the regional paediatric neuroscience centres participating in neuroscience conferences, case discussion with peers, education, distance learning, and so on. The centres should be fully supported with facilities for investigation (e.g. paediatric-trained clinical measurement scientists in EEG; MRI under anaesthetic/sedation). Children should not have to travel to Dublin for these investigations. Outreach clinics would allow ease of access for the family, be informative and educational for the local consultant paediatrician and MDT, and allow integrated care in the community.

These projections for consultant requirements for the managed clinical network in paediatric neurology are for the next five years (2015-2020) as outlined in the original needs assessment document in 2007 (King et al., 2007). The numbers quoted are the absolute minimum and are well below international recommendations. However, they reflect the reality of what is achievable in such a five-year period given the difficulties recruiting consultant staff to the paediatric neurology service. It is anticipated that these services, once established, will expand over

time, bringing the number of consultant neurologists in line with international standards. The development, implementation and effectiveness of the managed clinical network in paediatric neurology are dependent on the expansion of hospital general paediatric and community paediatric services.

Care Pathway in Managed Clinical Network

Child with "seizure":



Neurology Team Members

The diversity of the disorders managed by paediatric neurology services means that the MDT is large. Different members of the MDT will be more or less involved, depending on the disorder of the individual patient. An integrated paediatric neurology service should have at a minimum:

Consultant paediatric neurologist 1-2 per 100,000 children

Consultant clinical neurophysiologist 1:2,000 investigations (with four clinical neurophysiological measurement scientists and one administrator per consultant)

Multidisciplinary support team including:

- administration
- CNS/ANP
- speech and language therapist
- occupational therapist
- physiotherapist
- clinical psychologist
- neuropsychologist
- educational psychologist
- dietitian
- social worker
- play therapist
- music therapist
- access to inpatient teacher and school
- non-consultant hospital doctors (NCHDs) – one senior house officer (SHO) and one registrar per consultant (depending on rota) to ensure exposure of general practice and general paediatric trainees to paediatric neurology
- trainees in paediatric neurology possibly linked to the Royal College of Paediatrics and Child Health (RCPCH) paediatric neurology training programme

Specific support of:

- neurosurgery (paediatric) on site
- neuropathology
- neuroradiology (including interventional radiology, thrombectomy, embolisation)
- neurointensive care
- anaesthesia

Access to general paediatrics and paediatric surgery plus the full range of paediatric specialities including:

- respiratory
- cardiology
- endocrinology
- palliative care
- orthopaedics
- metabolic medicine
- psychiatry
- genetics
- neurorehabilitation, including acute rehabilitation – these services need multidisciplinary input of a highly specialised nature coupled with good liaison with local teams on discharge, as part of agreed care pathways in clinical networks.
- ophthalmology
- ENT/audiology

Well-developed neurodisability service in the community (physical and learning disabilities)

Transition to adult services, for example in epilepsy (advice regarding pregnancy, contraception, drug interactions and genetic counselling) and in neuromuscular disorders, especially those with life-limiting disorders.

Managed clinical networks for common disorders, e.g. epilepsy

Education – initial and ongoing education for all involved in childhood epilepsy, for example, is vital in order to develop and maintain expertise.

Facility for research

Databases on all children with neurological disorders to facilitate epidemiological analysis, research and audit

34.4 REQUIREMENTS FOR SUCCESSFUL IMPLEMENTATION OF MODEL OF CARE

Staffing

The minimum requirements for an integrated paediatric neurology service have been outlined in the model of care above. Once again, taking epilepsy as an example, the roles of the members of the MDT are outlined below. The same principles apply to other less common disorders such as neuromuscular disease, demyelination, movement disorders, stroke, and narcolepsy.

Role of secondary care nurses

- Coordinate local epilepsy working group, including education, learning disability, social services, school nurses, mental health services, parents and children
- Counselling
- Source of validated information
- Family support/child guidance liaison
- GP liaison
- Medication check/dosage adjustment

Role of secondary care paediatrician with responsibility for epilepsy

- Maintain standards of diagnosis and treatment and appropriate referral patterns along the clinical pathway
- Liaison with district-based colleagues over selection of children requiring referral to tertiary-based services
- Attend specialist epilepsy outreach clinics
- Regular attendance at regionally based special interest groups in epilepsy
- Supervision of care of children with intractable epilepsy
- Audit and improve local service delivery

Role of ANP in paediatric neurology/epilepsy

- Education and support of children, and families of children, with epilepsy
- Chronic disease management for refractory disease including transitional clinics to adult neurology
- Rapid access clinics (for ED discharges and first seizure)
- Telephone advice and email support for patients and families

Role of ANPs – Ambulatory and community role in epilepsy

- Support of inpatients, families and parents of children admitted with seizures
- Support/liaison of patients and families on the epilepsy surgery programme
- Every child with epilepsy in Ireland will have a named ANP attached to one of the paediatric centres. Patients attending services outside of their regions will be encouraged to transfer care back to the regional centre so as to avail of the ANP service. The nurse-led service will be a protocol-driven, chronic disease model of care covering the following responsibilities:
 - rapid access clinics in acute hospitals providing acute neurological care
 - outreach clinics for intellectual disability services and non-acute hospitals
- GP support
- Liaison with regional paediatricians
- Telephone/email/web support for patients
- Links to Epilepsy Ireland (previously Brainwave – the Irish Epilepsy Association) and the Irish Branch of the International League Against Epilepsy (ILAE)
- Implementation of standard operating procedures and management operating procedures
- Registration and measurement of key performance indicators, quality outcomes and clinical audit
- Health services research, population health and clinical and translational research

Roles of health and social care professionals

Access to clinical or educational psychology, neuropsychology, dietetics, OT, physiotherapy and SLT services will be provided, as appropriate to patient need, particularly for those with complex epilepsy who may have significant co-morbidities. The integration of therapy services across the hospital service, primary care and intellectual disability services will be facilitated by the GP.

- Clinical psychology is involved in assessment and support of a child with chronic epilepsy, particularly adolescents, where group therapy is effective and plays an important role in diagnosis and treatment of the child with functional seizures.
- Educational psychology is involved in assessment and liaison with schools.
- Neuropsychology is involved in assessment of a child with complex epilepsy, particularly those being evaluated for epilepsy surgery, neurocognitive regression, and so on.
- Occupational therapists have a key role in assessment, treatment and discharge planning and rehabilitation of patients with complex neurological presentations such as epilepsy, and in the management of cognitive and safety issues including provision of advice and equipment for home adaptations and the use of assistive technology.
- Physiotherapy management of children with epilepsy is focused on assessment and intervention in relation to physical disabilities, e.g. cerebral palsy due to malformation or acquired injury, falls prevention, respiratory intervention in the acute phase, and subsequent rehabilitation.
- Patients, particularly those with complex epilepsy, may require SLT for a wide range of issues including epileptic aphasia, complex language disorder, dysarthrias, feeding problems, and pre-surgical evaluation.
- Children with intractable epilepsy can be considered for specialised dietary treatment which involves a ketogenic or very low carbohydrate diet. This diet is complex and requires the input of a specialised dietitian working with highly motivated parents. Efficacy of the ketogenic diet for the management of paediatric refractory epilepsy has been demonstrated in a number of randomised controlled studies, systematic reviews and meta-analysis.
- Clinical measurement scientists (formerly EEG technicians) are now considered intrinsic to the delivery of quality care to people with epilepsy. Their role in the national programme is to promote timely access to high-quality diagnostic neurophysiological tests, across Ireland, in an equitable and efficient manner.
- Medical social worker involvement is vital from first contact with the service to ensure that the family is receiving appropriate benefits and to provide support.
- Music therapy has a role in treatment and discharge planning, using music in the context of a therapeutic relationship to address emotional and social needs.

Roles of administrative staff

- Reassurance to the family as the first point of contact
- Coordination of appointments for outpatient and inpatient service
- Typing of inpatient discharge summaries and outpatient clinic letters
- Maintenance of database on patients attending the service
- Maintenance of statistics on inpatient and outpatient attendances and activity in line with HIPE, among others.

Infrastructure

Inpatients should be located in a specialist neurology ward with neurodisability/neurosurgery specialties, if applicable. This will ensure the maintenance and development of skills and expertise for all professionals caring for the patient.

The facilities required for paediatric neurology outpatient service should include close proximity to other members of the MDT and support staff (see above). Ideally, the multidisciplinary neurology and clinical neurophysiology teams should be located together to facilitate ease of access for the family and accessibility to the neurophysiology team for discussion of acute and complex cases.

Education and Training

It is recognised that there is a link between well-planned clinical services and successful research and education programmes, and that effective planning of services will support academic development which will, in turn, help to drive clinical excellence. This is particularly applicable to neuroscience, where the cause of many of the diseases is only just being determined. Identifying the cause(s) of disease(s) will, in due course, facilitate moving forward with intervention studies, and neurologists need to be in the position to readily do this when required.

For example, the British Paediatric Neurology Association (BPNA) has initiated a national educational programme of:

1. Paediatric epilepsy training (PET) at three levels. PET 1 (level 1) is aimed at helping professionals who anticipate clinical contact with children with suspected epileptic seizures. PET 2 (level 2) is for paediatricians and specialist nurses who will provide clinical management and are developing further expertise in epilepsy. PET 3 (level 3) is for those with tertiary/quaternary-level responsibilities. These standardised courses are being rolled out nationwide in the UK and Ireland, allowing participants to obtain appropriate and continuing training.
2. Headache
3. Movement disorders
4. Neonatal neurology

Nursing Education and Training

The creation of a new cohort of expertise at ANP level is supported. Professional accountability of nurses and midwives is clearly defined by the Scope of Nursing and Midwifery Practice Framework (2000). Each nurse and midwife is individually accountable for his/her professional practice, including appropriate delegation. It is expected that new staff will be recruited at CNS or clinical nurse manager (CNM) 2 level. Defined competencies using a competency framework will allow these newly appointed nurses to move towards ANP over a predetermined period of time. The programme will commence with a post-registration course, which will cover basic physiological, pharmacological, psychological and social issues to do with epilepsy. All new nurses recruited will be expected to become registered nurse prescribers. A personal development plan, the competency framework with supported mentorship, benchmarking and extensive clinical experience will enable the novice epilepsy nurse, over time, to practise at an advanced level (Royal College of Nursing, 2013).

The Royal College of Surgeons in Ireland (RCSI) will deliver a master's programme in epilepsy nursing (inclusive of nurse prescribing), providing educational requirements to allow nurses to achieve the competencies necessary for registration as an ANP.

Medical Education and Training

A competent workforce is required at every level of the clinical network if care is to be delivered in an equitable way to all children and young people with neurological disorders and neurodisabilities. In the UK, there are currently national training grid programmes in paediatric neurology and paediatric neurodisability recognised by the RCPCH and the General Medical Council Medical Education and Training Board (GMC-METB). There is the potential to link the current training programmes more formally, in line with training programmes in other European countries, which would give a broader range of options in terms of future workforce development. This is currently being explored at the European Paediatric Neurology Society.

In 2004, the RCPCH approved the neurology departments in Crumlin and Temple Street for training in paediatric neurology, and this was confirmed in 2012. It is anticipated that the paediatric neurology unit in Cork University Hospital will also be approved for training in paediatric neurology by the RCPCH and will participate in a national training programme in paediatric neurology.

In addition to the above options for national grid/equivalent training in paediatric neurology and paediatric neurodisability, the RCPCH encourages those individuals with aptitude to train as general paediatricians 'with expertise'. There are currently approved training programmes for those wishing to train with an expertise in epilepsy and another option in neurodisability. In addition to the fellowship programme, there will be an ongoing programme of education for the following groups:

- specialist registrars (SpRs) and non-SpR registrars in paediatric neurology
- medical students
- GPs

Research

There is a need to expand academic paediatric neurology, neurodisability and evidence-based practice. Research in clinical neuroscience needs to be maintained and developed in order to foster excellence in delivery of care. A critical mass of manpower and expertise is required to support effective clinical research, as well as access to sufficient numbers of affected patients, in order to answer research questions in a cost-effective way, usually within a timeframe of not more than three years.

While paediatric neurologists have held posts as 'chair in paediatrics' there is no academic chair in paediatric neurology in Ireland, and this situation should be addressed. There is a unique opportunity to create a world-class clinical and research centre via the network established with the new children's hospital, similar to that established at George Washington University (Packer and Zechman, 2011) and elsewhere.

34.5 PROGRAMME METRICS AND EVALUATION

Key Service Outcomes in Paediatric Neurology

- Reduce outpatient paediatric neurology waiting times to <6 months.
- Reduce outpatient attendance figures to 10 patients per three-hour clinic.
- Reduce travel times to paediatric neurology outpatient visits to <2 hours.
- Meet NICE guidelines in the management of a child presenting with first afebrile seizure.
- Improve life expectancy and quality of life in children with Duchenne Muscular Dystrophy and other progressive neurological disorders.
- Establish a higher specialist training programme in paediatric neurology.
- Appoint a chair in paediatric neurology, leading to development of a meaningful research strategy.

34.6 GOVERNANCE

The governance structure should include a lead clinician for paediatric neurology. He/she would represent the specialty as part of the new governance structure of the new children's hospital with four clinical directorates. The lead clinician would be responsible for:

- strategic direction with regard to a department of neuroscience
- quality improvement and ongoing audit
- criteria for referral
- coordination of outreach clinics
- key performance indicators
- reporting to group and hospital clinical directors

34.7 PATIENT AND FAMILY EXPERIENCE OF THE SERVICE

- Regular and formal contact with patient representative groups to ensure that a child-centred and family-centred service is supported.
- The Neurological Alliance of Ireland (NAI) is the national umbrella organisation for over 30 not-for-profit groups representing families with a neurological disorder, and it has had a substantial positive impact on care. In many childhood disorders, such as epilepsy, muscular dystrophy and cerebral palsy, the specific support groups have provided services, funded staff and high-quality research, in addition to campaigning for research and therapeutic trials. The NAI has made a submission (Neurological Alliance of Ireland (NAI), 2015) to the neurology model of care.
- The 'Epilepsy12 Round 2 National Reports' (RCPCH, 2014; RCPCH, 2014) are an excellent example of the benefits of consultation with, and feedback from, users of a clinical network.
- The issue of lengthy travel is an important one and the development of managed clinical networks with outreach to paediatric departments is a very significant development. These outreach clinics will be paid for by the relevant hospital groups and will take place at a frequency that is sustainable (usually 2-3 monthly) with the local paediatrician and MDT obliged to co-attend.
- Many children with neurological disorders require lengthy hospital admissions for investigation and treatment. Families should be provided with adequate accommodation and facilities in or near the hospital during this very stressful period.

34.8 KEY RECOMMENDATIONS

- A managed clinical network for neurology should be implemented in Ireland, with outreach to regional paediatric centres, bringing together key professionals, and establishing well-defined integrated care pathways and clinical guidelines for each step of the pathway.
- Deficits in consultant paediatric neurologist posts, neurophysiology posts, and essential support services need to be addressed.
- An academic chair in paediatric neurology should be appointed.
- A training programme for consultants in paediatric neurology should be developed.

34.9 ABBREVIATIONS AND ACRONYMS

ADEM	acute disseminated encephalomyelitis
AEDs	anti-epileptic drugs
ANP	advanced nurse practitioner
BPNA	British Paediatric Neurology Association
CAMHS	Child and Adolescent Mental Health Services
CNM	clinical nurse manager
CNS	clinical nurse specialist
CSF	cerebrospinal fluid
CT	computerised tomography
ECG	echocardiogram
ED	emergency department
EDS	excessive daytime sleepiness
EEG	electroencephalography
EMG	electromyography
ERG	electroretinography
ESES	electrical status epilepticus in sleep
GMC-METB	General Medical Council Medical Education and Training Board
GP	general practitioner
GTCS	generalised tonic-clonic seizure
HIPE	Hospital Inpatient Enquiry
HLA	human leukocyte antigen
HSE	Health Service Executive
HSPC	Health Protection Surveillance Centre
ILAE	International League Against Epilepsy
IV	intravenous
MDT	multidisciplinary team
MRI	magnetic resonance imaging
MR	magnetic resonance
MSLT	Multiple Sleep Latency Test
NAI	Neurological Alliance of Ireland
NCHD	non-consultant hospital doctor
NHS	National Health Service
NICE	National Institute for Health and Care Excellence
NIOM	neuro-intraoperative monitoring
OT	occupational therapy
PET	positron emission tomography
PET	paediatric epilepsy training
PICU	paediatric intensive care unit
PSG	polysomnography
RCPCH	Royal College of Paediatrics and Child Health
RCSI	Royal College of Surgeons in Ireland
REM	rapid eye movement

SHO	senior house officer
SIGN	Scottish Intercollegiate Guidelines Network
SLT	speech and language therapy
SOREMP	sleep onset REM period
SPECT	single photon emission computed tomography
SpR	specialist registrar
UK	United Kingdom
WTE	whole-time equivalent

34.10 REFERENCES

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34.11 APPENDICES

34.11.1 Appendix 1 Disorders Seen by Paediatric Neurology Services

Epilepsy and other paroxysmal disorders

The largest single patient group covered by paediatric neurology services is patients with epilepsy (prevalence 0.7/100). However, the majority of patients with epilepsy are wholly managed either by secondary care providers, or by community-based paediatricians, sometimes with advice from paediatric neurology services. The role of paediatric neurology services in the management of children with epilepsy is defined by NICE guidelines. These indicate the need for paediatric neurology services in epilepsy beginning under the age of two years; epilepsy accompanied by other neurodevelopmental problems, such as learning difficulties; epilepsy associated with abnormal brain imaging; in cases of diagnostic doubt; and epilepsy not responding to appropriate first-line anti-epileptic drugs (AEDs). The guidelines also state that children and young people with recent onset suspected seizure should be seen urgently by a specialist as part of the paediatric neuroscience network service. The role of paediatric neurology services includes the use of newer AEDs, the use of non-drug treatments such as the ketogenic diet, and the selection of patients for surgical treatment of epilepsy, vagal nerve stimulation and novel treatments.

Acute encephalopathies

These disorders include coma (non-traumatic coma incidence approximately 50/100,000/year), refractory status epilepticus, infectious and inflammatory diseases of the nervous system, including complicated meningitis and encephalitis, and other causes of acute non-traumatic encephalopathy (e.g. acute disseminated encephalomyelitis (ADEM)).

Some children with these disorders are managed by secondary-level paediatric services.

Demyelinating disorders

These include multiple sclerosis, clinically isolated syndrome, optic neuropathy, neuromyelitis optica, among others. These disorders are rare and are managed on an individual basis, often in consultation with adult neurology colleagues and the special interest group of the BPNA on neuroinflammation and infection.

Cerebrovascular disorders (including childhood stroke)

These disorders are managed by paediatric neurology services which, working with neuroradiology and paediatric neurosurgery, are responsible for acute management, investigation of cause and referral for subsequent rehabilitation. Most acute stroke in children without cardiac/haemo-oncology disease is inflammatory in aetiology. At present there are no international guidelines on the use of thrombolysis in acute stroke in children. Within the BPNA there is a special interest group on cerebrovascular disease.

Acute paralysis

This includes transverse myelitis, Guillain-Barré syndrome and spinal cord lesions. Many of these children will require acute spinal/brain imaging and are referred to the paediatric neurology service.

Motor disorders (including unexplained cerebral palsy, ataxia, and movement disorders)

Paediatric neurology services are principally involved in diagnosis and investigating underlying causes. While movement disorders are managed by paediatric neurology, therapeutically, many children with cerebral palsy are managed by neurodisability services. Paediatric neurology services may provide specialist spasticity management, usually working in partnership with disability orthopaedic and neurosurgery services

Neuromuscular disorders

These include congenital myopathies, muscular dystrophies (Duchenne prevalence 0.3/1,000 male births), spinal muscular atrophies and neuropathies. Paediatric neurology services are involved in diagnosis and, in collaboration with disability services, for long-term management including new treatment trials. Management of complications requires collaboration with spinal, respiratory (including non-invasive ventilation) and cardiac services. End-of-life care is an important aspect of services for some of these disorders. (See Appendix 7)

Brain and spinal cord tumours

Brain tumours are the most common solid malignancies in childhood, and four per 100,000 children aged 0-16 years will be diagnosed with a tumour of the central nervous system. The MDT responsible for diagnosing and managing children includes paediatric oncology, paediatric neurology and paediatric neurosurgery, in addition to long-term support from other health and social care professionals.

Developmental delay, learning and behavioural difficulties, including autistic spectrum disorders

Paediatric neurology services are mainly involved in investigating the underlying cause of these disorders. Long-term management is usually undertaken by community services and CAMHS.

Headache, including migraine

Headache is the most common neurological disorder in childhood. It is mainly managed in primary and secondary care. However, paediatric neurology services are involved in diagnosis and in managing a minority of children with complex headache syndromes, including migraine not responding to first-line treatments, chronic daily (tension) headache and idiopathic intracranial hypertension.

Neurogenetic disorders, neurocutaneous syndromes and dysmorphic syndromes involving the nervous system

Paediatric neurology is involved in diagnosis and management of epilepsy, if present, and referral for genetic counselling, if relevant.

Central nervous system malformations and spinal dysraphism

Paediatric neurology is involved in diagnosis and management of epilepsy, if present.

Functional disorders

The treatment of children with functional disorders requires the involvement of paediatric neurologists in diagnosis and in advising on rehabilitation with an MDT that includes psychiatric input.

Traumatic brain injury, including non-accidental head injury

Paediatric neurology services generally provide advice regarding specific aspects of medical management as part of an MDT also involving various surgical disciplines. Diagnosis of suspected non-accidental head injury involves a multidisciplinary and multi-agency approach.

Neonatal neurological disorders including seizures, hypotonia, stiffness, and dysmorphism

Paediatric neurology may be involved in diagnosis and management in collaboration with the neonatal team.

Neurometabolic and neurodegenerative disorders

This is a large group of mostly rare conditions. Paediatric neurology services are involved in diagnosis and sometimes in long-term management. Recent advances in genetic therapies indicate that survival will improve in children with some degenerative disorders.

Neurological manifestations of systemic disease

Examples include autoimmune, renal, and metabolic disorders. Paediatric neurology is involved, usually in consultation with a primary specialist team.

Severe behavioural/psychiatric disorders in children who may have neurological symptoms

Paediatric neurology is involved in assessment and exclusion of neurological disorders.

34.11.2 Appendix 2 Conditions Excluded from Paediatric Neurology Services

In general, children with certain neurological disorders will be excluded from the paediatric neurology service and will be seen/managed in secondary care or by other specialist services. These disorders, and the disciplines by which they are mainly or often managed, are:

- 'Simple' headache – primary care and secondary paediatrics
- Febrile seizures – secondary paediatrics
- Vasovagal (syncope); breath-holding attacks; reflex anoxic seizures; infantile gratification – primary or secondary paediatrics
- Meningitis without significant neurological complications – general paediatrics
- Some types of epilepsy as defined in relevant NICE guidelines – secondary paediatrics
- Learning problems – neurodisability. However, the investigation of the cause of learning problems is often the responsibility of paediatric neurology services.
- Behavioural problems and social and communication disorders, including autistic spectrum disorders – secondary paediatrics, neurodisability, CAMHS. However, the investigation of the cause of these problems is often under paediatric neurology services.
- School difficulties – community/general paediatrics
- Dyspraxia – patients may be seen and diagnosed by occupational therapy and general/community paediatric services; they will be referred to neurology only if other abnormal features are present on history or examination by the paediatrician. A paediatric neurology assessment should not be required in order for a child to access services in the community
- Tics – most are seen in secondary paediatrics or CAMHS unless there are specific neurological signs or atypical features.

34.11.3 Appendix 3 Neurophysiology Requests

EEG referrals

- Referrals for routine and sleep EEG studies are accepted from paediatric neurologists, paediatricians, physicians practising in intellectual disability services, and psychiatrists.
- Requests for video EEG telemetry (vEEG) are only accepted from paediatric neurologists or epileptologists. If a paediatrician considers that a vEEG might be indicated, it is best to discuss the case with the paediatric neurologist or epileptologist, who may need to see the patient prior to making a decision. Please refer to indications for vEEG below.
- Patients should be referred to the department closest to home, and to which the referring doctor plans to refer the child for neurological opinion, should this be necessary. This avoids duplication of services and unnecessary re-evaluation of tests such as EEG studies and imaging.
- Communication between the neurophysiologist/epileptologist and paediatricians regarding urgent or semi-urgent EEGs should be encouraged, and consultant paediatricians at secondary care centres should have access to the consultant neurophysiologist/epileptologist to advise on the degree of urgency of EEG.
- EEG results: Formal reports will be sent to the referring physician. The consultant neurophysiologist/epileptologist will identify EEG results requiring immediate action and will notify the referring physician.

Urgent or soon EEGs

1. **Urgent:** There are few indications for urgent EEG studies. These include the suspicion of ongoing non-convulsive status epilepticus or unexplained presentation of encephalopathy/depressed sensorium without a clear cause.
2. **Very soon** (within a few days): This applies to few patients, and the most frequent or obvious indication is suspicion of infantile spasms. At the moment, and for this particular indication, it is imperative that a home video or hospital video be taken and reviewed. This may resolve the problem by confirming the likelihood of the diagnosis clinically and expediting the EEG or, indeed, excluding the diagnosis. This is very helpful, particularly if the question arises during weekends or holidays with minimal staff cover.
3. **Soon:** This is a situation where the neurologist/epileptologist needs to make a judgement as to why and how soon an EEG is needed. Examples of such situations are:
 - a. Suspicion of absence seizures (frequency is paramount, in order for that suspicion to warrant a soon EEG)
 - b. Explosive onset of frequent seizures of any type where clinical judgement inclines one to start treatment with AEDs with no delay
 - c. Escalation or rapid evolution of an epilepsy presentation while awaiting a routine EEG (i.e. seizures becoming very frequent after a slow/infrequent presentation or onset or emergence of several generalised tonic-clonic seizures (GTCSs) which may warrant initiation of AED treatment). In the latter two cases, it is very likely that these patients will be referred to neurology either for advice on treatment or inpatient management.

Appropriate and inappropriate referrals for EEG studies

Appropriate

- i. definite, probable seizure/epilepsy
- ii. classification of newly diagnosed epilepsy
- iii. established epilepsy with suspicion of sub-clinical seizures/ frequency
- iv. established epilepsy with change in semiology (from focal to generalised or vice versa provided there is no known structural abnormality)
- v. suspicion of non-convulsive status epilepticus (either de novo, or after treatment of convulsive status epilepticus)
- vi. work-up for encephalopathy/regression/developmental delay (e.g. suspected Angelman syndrome, where EEG may facilitate early diagnosis)
- vii. weaning AEDs: only in patients with absence epilepsy or previously refractory epilepsy including the epileptic encephalopathies (e.g. spasms)
- viii. repeat EEGs to monitor treatment in epileptic encephalopathy (preferably with sleep)

Inappropriate

- i. funny turns when there is a clear history of breath-holding/tics/vasovagal events; EEG is actually contraindicated in these cases (false positive findings)
- ii. drug withdrawal for well-established benign focal epilepsy
- iii. established epilepsy in patients with known substantial structural abnormalities – clinical change in seizure semiology (*)
- iv. febrile seizures (unless Dravet syndrome and its variants are suspected; prolonged repeated in first year in life)

- v. day-dreaming or bad behaviour in patients with autism (unless the paediatrician or neurologist suspects independent seizures)
- vi. prolonged visual or auditory hallucinations in psychiatric patients other than the well-described elemental hallucinations of parietal/temporal lobe epilepsy**

(*) *Example: patient with lissencephaly having GTCSs changing to tonic seizures or to more focal seizures or vice versa...an EEG will not help how you treat.*

(**) *These cases in particular should be discussed with a neurologist or epileptologist.*

Video-EEG Telemetry indications

- i. capturing and characterising clinical seizures to help establish and define/classify specific types of epilepsy and/or epilepsy syndromes
- ii. suspicion of non-convulsive/subclinical seizures or electrical status epilepticus in sleep (ESES) in children with known epilepsy and altered mental state or acute deterioration in cognition
- iii. determining seizure load throughout the day/night; for example, patients with subtle/brief absence or dyscognitive seizures which can often be missed by simple day-to-day observation
- iv. evaluation of patients with refractory epilepsy for potential epilepsy surgery
- v. capturing and characterising potential non-epileptic events

Requesting EMG and nerve conduction studies

- Nerve conduction studies and EMG are used to establish the presence of neuropathy, anterior horn cell disorders, neuromuscular transmission defects and myopathy.
- The studies are performed in a manner that minimises discomfort in children and are generally well tolerated.
- In almost all instances, request for EMG is a routine outpatient study. Rare situations where urgent EMG may be appropriate include severe acute, or rapidly evolving (within days) neuromuscular weakness, especially where bulbar or respiratory weakness may feature.

Appropriate (useful indications) for EMG/nerve conduction studies

- i. establish presence of, and characterise, (axonal versus demyelinating) peripheral neuropathy
- ii. delineate the distribution and examine recovery potential of peripheral nerve or plexus traumatic injury. (This can contribute to decisions regarding peripheral nerve surgery or exploration).
- iii. investigate myopathic weakness
- iv. investigate suspected mysathenic disorders in neonates or older children

Inappropriate (unhelpful indications) for EMG/nerve conduction studies

- i. investigation of weakness or hypotonia with a clearly 'central nervous system' origin
- ii. follow-up studies in established and biologically inactive neuropathy or myopathy syndromes or in known cases of myasthenia. (EMG is not a surrogate for clinical outcome measures in any of the above.)
- iii. evaluation of suspected axonal injuries within three weeks of onset of injury. (It takes up to three weeks for EMG features of acute axonal injury to appear; study inside of this window of time is too soon).

34.11.4 Appendix 4 Guidelines for Treatment of Status Epilepticus (for children >3 months)

Definition of status epilepticus

1. prolonged seizure activity (>5 minutes for GTCS)
2. recurrent seizures without recovery of consciousness

Take blood for:

- Glucose*, urea and electrolytes, calcium, magnesium, blood gas, full blood count, blood cultures
- AED levels, toxicology screen (as appropriate)

* If low blood sugar <2.6mmol/L administer dextrose 10% 2mg/kilogramme and re-check sugar in 10 minutes.

* If patient is on ketogenic diet, refer to protocol on ketogenic diet before prescribing intravenous (IV) fluids (i.e. dextrose) and/or consultant neurologist on call.

Check if any rescue medications were administered before presentation to hospital.

Start AED treatment:

1. First Line: Benzodiazepines

The preferred benzodiazepine is Lorazepam because of a much longer duration of action.

1. Lorazepam (Ativan®)

Intravenous bolus: 0.1mg/kg (maximum dose= 4mg)

Given as a slow push

Dilute with an equal volume of sodium chloride 0.9%.

2. Buccal Midazolam (Buccolam®)

Can administer while obtaining IV access

Dose is age dependent

3 months-1year: 2.5mg

1-5years: 5mg

5-10 years: 7.5mg

10-18 years: 10mg

3. Diazepam (Stesolid®)

Has equal onset to IV Lorazepam, but shorter duration of action

Can administer while obtaining IV access

Dose is age dependent

1 month-2 years: 5mg

2-12 years: 5-10mg

12-18 years: 10-20mg

The above step may be repeated (two doses in total) if ongoing seizures 10 minutes after the first dose of benzodiazepine. Check if parents/school/ambulance have administered benzodiazepines before presentation.

After 10 minutes, move to second line.

2. Second Line: Fosphenytoin

Measured as phenytoin sodium equivalents (PE units)

Loading dose: 20mg PE/kg by IV infusion

Rate of infusion: 3mg/kg/minute

Maintenance dose: 2.5mg/kg/dose 12-hourly

To be commenced 12 hours after loading dose

If no Fosphenytoin Sodium available, can use Phenytoin Sodium

Loading dose: 20mg/kg IV

Rate of infusion: 1mg/kg/minute

Maintenance dose: 2.5mg/kg/dose 12-hourly

After 10 minutes: contact medical consultant on call and move to third line.

3. Third Line: Other Aeds For Treatment Of Status Epilepticus

1. A. Levetiracetam (Keppra®)

IV loading dose: 20-40mg/kg

Maximum dose: 1,500mg

Dilute in at least 100ml of 0.9% sodium chloride or dextrose 5%

Rate of infusion: 15 minutes

B. Lacosamide (Vimpat®)

IV loading dose: age dependent

< 2 years: 50mg

2-8 years: 100mg

>8 years: 200mg

Can give undiluted

2. Sodium Valproate (Epilim®)

DO NOT USE for fir presentation of seizures in children <2 years or children with suspected metabolic condition or an underlying liver condition.

IV loading dose: 10mg/kg

May dilute in 0.9% sodium chloride or dextrose 5%

Give over 3-5 minutes.

After 10 minutes, move to fourth line.

4: Fourth Line: Phenobarbitone

IV loading dose: 10-20mg/kg

Dilute with water for injection to concentration of 20mg/mL

Slow intravenous injection, no faster than 1mg/kg/minute

Maintenance 2.5mg/kg/dose 12-hourly

Drug levels are important in monitoring treatment.

5: Fifth Line: Contact Icu

Midazolam infusion or Thiopentone

Midazolam infusion

Dose: 1-4 micrograms/kg/min

Dilute 3mg/kg in 50mls of 0.9% sodium chloride or dextrose 5% for final concentration of 1ml/hour = 1mcg/kg/min

Thiopentone infusion

Dosing as per PICU protocols.

Guidelines for preparation and administration of Fosphenytoin (Pro-Epanutin®) loading doses

- Measured as phenytoin sodium equivalent (PE)
- Loading dose = 20mg (PE)/kg/IV
- Rate of infusion = 3mg (PE)/kg/minute = maximum 50mg (PE)/minute
- Must be administered with IV syringe pump only
- Concentration range: 1.5-25mg(PE)/ml
- **DO NOT USE PRO-EPANUTIN®UNDILUTED: Following dilution, use immediately**

Estimated weight (Kg)	Dose of Fosphenytoin (Mg (PE))	Volume of Fosphenytoin solution required (50mg (PE) in 1ml) (mL)	Volume of dextrose 5% or 0.9% sodium chloride to be ADDED to Fosphenytoin (mL)	Total volume (Fosphenytoin PLUS diluent) (mL)	Infusion time (minutes)
3	60	1.2	5	6.2	7
4	80	1.6	5	6.6	7
5	100	2	5	7	7
6	120	2.4	5	7.4	7
7	140	2.8	5	7.8	7
8	160	3.2	5	8.2	7
9	180	3.6	5	8.6	7
10	200	4	10	14	7
11	220	4.4	10	14.4	7
12	240	4.8	10	14.8	7
13	260	5.2	10	15.2	7
14	280	5.6	15	20.6	7
15	300	6	15	21	7
20	400	8	15	23	8
25	500	10	20	30	10
30	600	12	20	32	12
35	700	14	20	34	14
40	800	16	25	41	16
45	900	18	25	43	18
50	1,000	20	25	45	20
55	1,100	22	30	52	22
60	1,200	24	30	54	24
65	1,300	26	30	56	26
70	1,400	28	30	58	28

Use 20ml Syringe*

Use 50ml Syringe*

*B. Braun Perfusor syringe pump –

o 20ml syringe – Max rate 100ml/hr

o 50ml syringe – Max rate 200ml/hr

Please note that the above guide is for loading doses only

Department of Neurology and Clinical Neurophysiology

Temple Street Children's University Hospital Checked by M. Kirrane/R. Patel Pharmacy Department, TSCUH

Updated September 2015

34.11.5 Appendix 5 Guidelines for treatment of status epilepticus (for infants <3 months)

First line	Phenobarbitone Loading dose: 20mg/kg IV Can repeat loading dose (maximum dose 40mg/kg IV) Maintenance dose: 2.5mg/kg/dose IV 12-hourly
↓	
10 Minutes	
Second line	Fosphenytoin Measure as phenytoin sodium equivalents (PE units) Loading dose: 20mg (PE units)/kg IV Or Phenytoin sodium Loading dose: 20mg/kg IV
↓	
10 minutes – call medical/ED consultant	
Third line	Levetiracetam (Keppra®) Loading dose: 20-40mg/kg IV
↓	
Fourth line	Further treatment after contacting consultant neonatologist on call/consultant neurologist on call

34.11.6 Appendix 6 Out-of-hospital Rescue Medication

Indications for prescribing and giving buccal midazolam

A. Buccal midazolam indications (prescribing and administering) for patients in the community (message to GPs, schools)

- Always remember it is an EMERGENCY medication
 - It is not for shortening prolonged complex partial seizures which have a minimal motor or respiratory component.
1. Prolonged GTCS i.e. for more than five minutes (emergency)
 2. Tonic seizures with cyanosis or prolonged more than five minutes (emergency)
 3. Known patients who usually have rare or infrequent seizures, if they cluster frequently over a short period on one day and/or are known to progress to habitually prolonged GTCSs when they cluster (preventing a very likely emergency)
 4. If a patient required emergency medication in the past, and is weaning off their anti-epileptic medications, they may have rescue medication for up to a further two years.

Situations in which buccal midazolam should NOT be prescribed

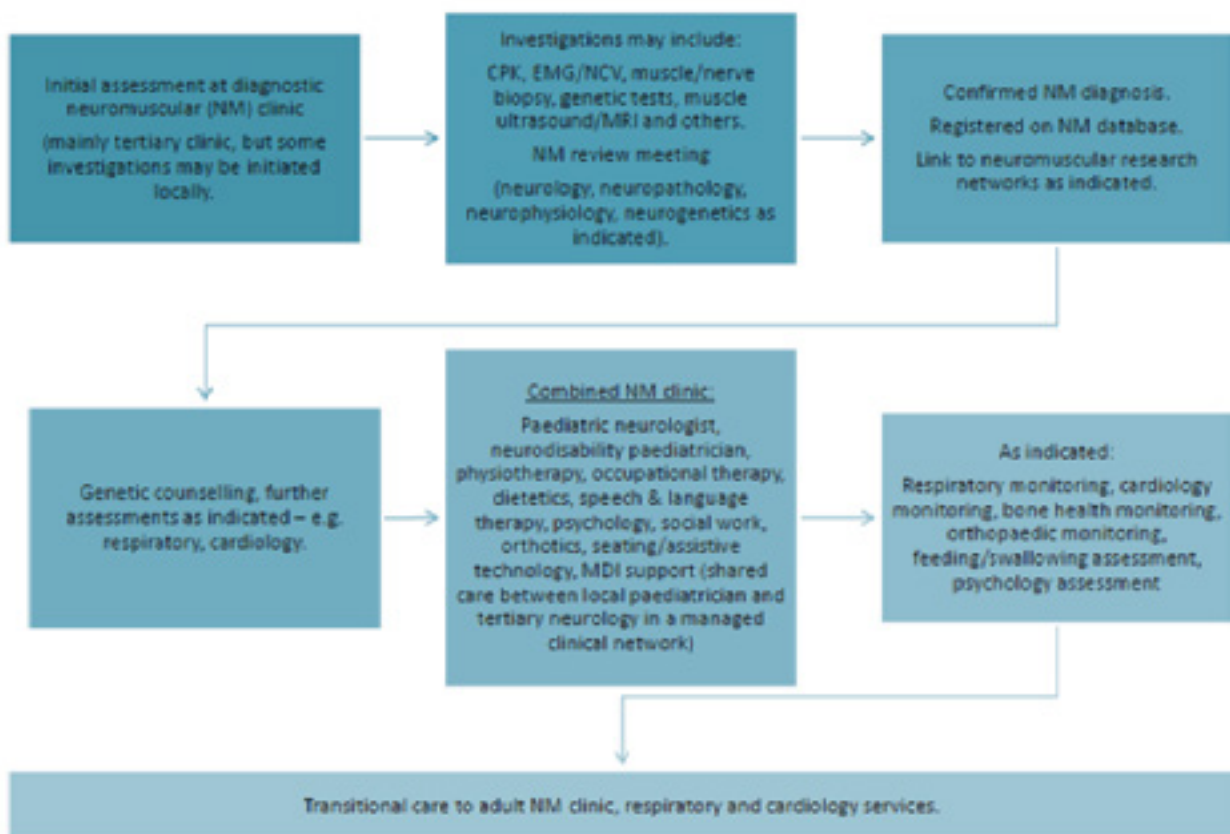
1. Occipital lobe seizures with no effect on breathing or cyanosis
2. Prolonged complex partial seizures which have a minimal motor or respiratory component
3. Requests from parents, schools or day-care institutions for children (who had it prescribed in the past, but have been seizure free for a year, particularly if it was not needed frequently in the past).

B. Buccal midazolam exceptional indications to prescribe/administer at the discretion of paediatric neurologists in charge of specific patients

1. Patients who live in remote areas with a known prolonged emergency/ambulance response time and who are known to have GTCSs, primary or secondary generalised.
2. Patients who live in remote areas with a known prolonged emergency/ambulance response time could be allowed a second dose of buccal midazolam if first dose has not terminated the seizures within 15-20 minutes of administration provided that it is continuing to escalate and spread with no signs of slowing down or fading, and provided that ambulance is called. This is particularly relevant and justified if it has been used before in such settings (twice within 24 hours) and is known to have been tolerated with no respiratory depression.

There are cases that need individualised evaluation and there are exceptional circumstances where a consultant at his/her discretion may prescribe this drug.

34.11.7 Appendix 7 Neuromuscular Model of Care



34.11.8 Appendix 8 Narcolepsy Model of Care

Narcolepsy – Brief overview

Introduction

Narcolepsy is characterised by four classic symptoms:

- excessive daytime sleepiness
- cataplexy
- hypnagogic hallucinations
- sleep paralysis

The aetiology of narcolepsy is still not fully understood. However, the strong association with human leukocyte antigen (HLA) genotype HLA-DQB1*0602 genotype and HLA-DR2 and HLA-DQ1 and some experimental evidence suggests an ‘autoimmune’ origin. This might reduce the number of neurons producing a protein called hypocretin, which is heavily involved in controlling sleep and appetite. There is currently an acceptance that the use of a specific H1N1 vaccine during the 2009/2010 swine flu epidemic was associated with subsequent development of symptoms of narcolepsy, particularly in young people. This has been investigated by the Health Protection Surveillance Centre (HSPC).

Clinical Features

‘Excessive daytime sleepiness’ (EDS) is the primary symptom and must be present for >3 months. Sleep occurs in difficult and potentially embarrassing or even life-threatening situations such as driving, eating or talking and may occur without warning – sleep attacks. Patients tend to take frequent naps during the day which can be associated with dreaming. A score of 10 or greater on the Epworth Sleepiness Scale suggests EDS that warrants investigation.

- ‘Cataplexy’ is a brief and sudden loss of tone because of sudden rapid eye movement (REM) intrusion. It is often associated with an emotional trigger such as laughter or anger. Respiratory and external ocular movements are usually functioning during cataplexy. Cataplexy is found in about 70% of patients with narcolepsy and its presence with EDS suggests that this is narcolepsy. However, up to 30% of patients with narcolepsy do not have cataplexy.
- ‘Sleep paralysis’ is the inability to move upon waking, or just after falling asleep, with consciousness intact. Again, respiratory and extraocular movements are usually spared.
- ‘Hypnagogic and hypnopompic hallucinations’ occur at sleep onset and waking, respectively. They are usually vivid and can have visual, auditory or tactile elements.

Symptoms or concerns that might flag a case of narcolepsy in children include obesity, deterioration in school performance, poor concentration, and emotional lability. Examination is generally normal. During cataplexy there is atonia and loss of deep tendon reflexes.

Diagnostic tests

HLA typing: This is generally clinically helpful to exclude narcolepsy if negative (since these alleles are found in up to 35% of the population).

CSF hypocretin levels: Levels <110 pg/mL are indicative of narcolepsy. However, higher levels do not exclude narcolepsy.

MRI brain scanning is generally normal. However, it can be useful to exclude lesions that might be causing secondary narcolepsy.

Neurophysiology testing: Polysomnography (PSG) and Multiple Sleep Latency Tests (MSLTs) are essential in the diagnosis. All central nervous system stimulants must be stopped for two weeks prior to testing. These tests help by (a) doing PSG to exclude other causes of EDS and (b) MSLTs can have specific features to suggest narcolepsy. The MSLT involves measuring five opportunities to nap at two-hour intervals during the day. The findings of more than two sleep-onset REM periods (SOREMPs) and a mean sleep latency of <8 minutes strongly suggest narcolepsy. However, interpretation can be difficult and some patients require repeat testing.

Narcolepsy – Information on drugs used

Central Nervous System Stimulants:

Daytime wakefulness

Methylphenidate – short acting 2-3 times daily: morning, lunch and 3-4pm		Methylphenidate – modified release Once daily morning +/- short-acting in the evening	
Brands	Strengths Maximum daily dose 60mg	Brands	Strengths max dose 60mg
*Ritalin	10mg	Concerta XL	18mg 27mg 36mg
*Medikinet	5mg 10mg 20mg	Equasym XL	10mg 20mg 30mg
		Medikinet MR	5mg 10mg 20mg 40mg
		Ritalin LA	20mg 30mg 40mg

* Prescription written in words and figures. Total amount to be dispensed to be documented. Max 90 day supply

Non-amphetamine wake promoter: Modafinil

Morning and lunch time

Brand	Strength - max dose 400mg
Provigil	100mg 200mg

Cataplexy treatment 5HT/NA reuptake inhibitor: Venlafaxine

Morning and lunch time

Brands	Strengths 37.5mg, 75mg, 150mg taken with stimulant medications. SR version at lunch time can aid with nightmares, sleep paralysis and/or hallucinations
Multiple brands see MIMS	

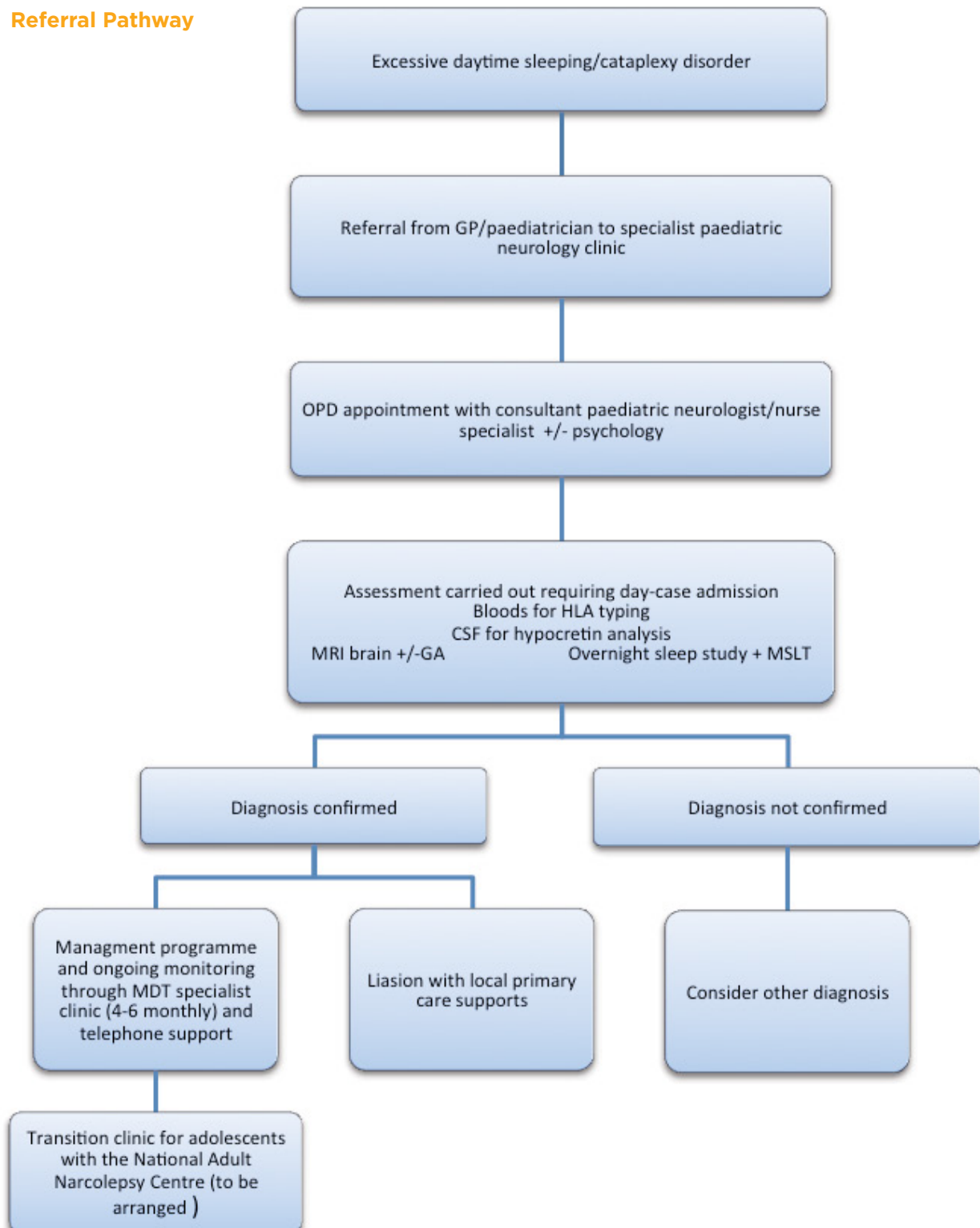
Central nervous system depressant – consolidates night-time sleep, promotes daytime wakefulness, cataplexy treatment: Sodium oxybate

Taken on going to bed and 2.5-4 hours later, on empty stomach

Brand	Strength starting dose 60-90mg/kg in two divided doses. Adult start dose 4.5 grammes divided dose. Max dose 180mg/kg or 9 grammes.
Xyrem	500mg/ml

High-tech prescription. Prescription written in words and figures, total amount per month to be dispensed must be documented.

Referral Pathway



34.11.9 Appendix 9 Guideline on Paediatric Headache

Recurrent headache in children is common, and migraine is the most common cause of severe recurrent headache in this group. Migraine affects almost 1:10 school children. The features in many children are similar to those in adults, but there may be differences.

Types of headache

The headache types seen typically in children presenting to general practice are, in descending order:

Frequency:	Type:
Very common	Migraine Tension type
Common	Sinusitis related Analgesic related
Uncommon	Systemic illness Cluster headaches
Rare	Idiopathic intracranial hypertension Intracranial space occupying lesion Systemic hypertension

Temporal features

- Acute progressive, i.e. rapidly increasing – e.g. intracranial hypertension, meningitis
- Acute recurrent, i.e. free in between attacks – migraine
- Chronic progressive, i.e. most days or several days a week slowly getting worse, often over months or years – e.g. raised intracranial pressure, tumours
- Chronic non-progressive, i.e. daily, rarely severe – emotional/psychological features may be important
- Mixed, i.e. constant daily, not severe (tension) + recurrent (not) daily episodes of acute severe (migraine).

Migraine headaches

- May be present with aura (migraine with aura) previously classified as classic, ophthalmic, hemi-paraesthetic, or hemiplegic migraine
- Frequently with prodrome, i.e. difficulty concentrating or autonomic features
- Migraine without aura (previously known as common migraine) where the headache is commonly bilateral and usually in the occipital area.

However, most children with migraine tend not to have typical syndromes, but can have bilateral or unilateral headaches typically with nausea and vomiting but with or without photophobia or phonophobia, blurred vision, diplopia or flashing lights (Newton, 2008).

Tension headaches

- Infrequent episodes of headache can last between minutes and days.

Other types of childhood headache syndromes (periodic syndromes)

- cyclical vomiting
- abdominal migraines
- benign paroxysmal vertigo

Cluster headaches (chronic paroxysmal hemicranias)

- These are uncommon in childhood, and are characterised by severe unilateral pain associated with the following: conjunctival injection, rhinorrhoea, lacrimation, meiosis or ptosis.

Both migraines and tension headaches can often be managed in the same way.

Table 1 – History and Physical Features

<p>History:</p> <p>Encourage child to use own words</p> <ul style="list-style-type: none"> • Date of onset • Frequency and severity • Duration of each attack • Site of maximal intensity • Any change in symptoms better or worse • Treatments used so far • Associated features, i.e visual deficits • Predisposing or trigger factors <ul style="list-style-type: none"> o Stress/anxiety, e.g. change in school o Menstruation o Oral contraceptives o Physical exertion/fatigue o Lack of sleep o Hunger o Foods/drinks with high amine content; caffeine or citrus drinks o Reading/refractive error o Cold foods 	<p>Physical features:</p> <p>Examination must include</p> <ul style="list-style-type: none"> • Blood pressure • Growth centiles • Any cutaneous signs re NF • Pubertal development (if relevant) • Neurological full exam, especially note: <ul style="list-style-type: none"> o Gait abnormalities o Coordination/tremor o Asymmetry o Fundoscopy o Visual fields o External ocular movements looking for nystagmus and diplopia. o Deep limb tendon reflexes (i.e. brisk and/or clonus) o Plantar responses
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When reviewing the history and completing a satisfactory exam (see Table 1) it is important to recognise any worrying features which require rapid referrals under the two-week rule.

Table 2: Worrying Features for Rapid Referral

<ul style="list-style-type: none"> • Night-time headache wakening child • Associated persistent vomiting or nausea (i.e. not episodic) • Deterioration in personality, schoolwork or behaviour • Recent change in character or quality of headache • Persistent and worsening headache • Headache worse when lying flat or with cough or straining 	<ul style="list-style-type: none"> • Recent onset squint • Signs of cerebellar dysfunction, i.e. ataxia • Features suggestive of raised ICP <ul style="list-style-type: none"> o Papilloedema o Diplopia o Nystagmus o Abnormally brisk deep tendon reflexes/clonus
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When to refer

This should be when the pattern is more suggestive of underlying pathology, or when treatment is ineffective.

Table 3: Indications for Referral

Diagnostic	Treatment	<2 weeks, i.e. two-week rule. Mark this on referral letter
<ol style="list-style-type: none"> 1. Seizures 2. Under four years 3. Anxiety 4. Unusual migraine 5. Any worrying features 	<ol style="list-style-type: none"> 1. Frequency affecting school attendance or otherwise disabling 2. Severe, psychological factors 	<p>Headaches of recent origin with one or more of the following features:</p> <ol style="list-style-type: none"> 1. Increasing frequency or severity, including nocturnal waking 2. Behaviour change or deterioration in school performance 3. Associated neurological signs

Table 4: Indications for Neuroimaging

<p>High priority</p> <p>Chronic progressive pattern Abnormal neurological exam Focal neurological symptoms Altered mental status Papilloedema Ataxia Abnormal deep tendon reflexes</p>	<p>Presence of VP shunt Presence of neurocutaneous syndrome Age under three years</p> <p>Headache or vomiting on awaking Seizures (present in 10% of migraine)</p>
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Treatment

1. General measures
 - a. Reassure
 - b. Identify and remove headache triggers
 - c. Regulate lifestyle (sleep and eating pattern)
 - d. Stress reduction
 - e. Dietary changes, i.e. low-amine diet (e.g. cheeses, Marmite, soft drinks).
2. Acute attack

Drugs should be given as early as possible and, in general, treatments should not be used for more than two days per week because of the risk of rebound headache (particularly with paracetamol).

 - a. Paracetamol (15mg/kg)
 - b. Ibrufen (10mg/kg)
 - c. Sumatriptan nasal spray (licensed for children aged 12 years and above only)

Evidence from randomised controlled trials for all of these drugs is limited, but paracetamol and ibrufen appear to be twice as effective as placebo. Evidence for effectiveness of nasal sumatriptan is more limited, with side effects increasing with the dose. Opioids (i.e. codeine) are not recommended.

- d. Antiemetics
 - i. Domperidone 0.25-0.5mg/kg three times/day (not licensed indication)
 - ii. Antihistamine antiemetics can also be used
 - iii. Avoid Metoclopramide
- 3. Preventing attacks

Both Pizotifen and propranolol are licensed for the treatment of migraine headaches in children.

Pizotifen

0.5mg tabs given at night. Side effects – weight gain, constipation, dry mouth, hyperactivity.

Propranolol

Licensed for treatment of migraine in children at a dose of 20mg two or three times a day for under-12s and 40mg three times a day for over-12s. Propranolol is contraindicated in asthma.

A recent systemic review showed that propranolol is more effective than placebo in the short term, but has unwanted side effects.

Antiepileptic agents

Sodium valproate (30mg/kg/day) and topiramate (4mg/kg/day) escalating over three weeks and continuing over six weeks once remission is obtained; this would normally be managed by a paediatrician with special interest in neurology or a neurologist.

The decision to start prophylaxis should be made together with the parent and child, with a discussion of side effects (antimuscarinic effects with weight gain for Pizotifen and tiredness, bronchospasm, sleep disturbance and hypotension with propranolol).

It is inappropriate to continue prophylaxis medication for long periods once attacks have subsided.

34.11.10 Appendix 10 Ketogenic Diet Referral Pathway

Sample Ketogenic Diet Pathway

Step 1	Step 2	Step 3	Step 4	Step 5
<ul style="list-style-type: none"> Referral: Referral from neurology consultant to neurology CNS and ketogenic dietitian Information sent to parents CNS and dietitian discuss referral First meeting with parents Dietitian and CNS have follow-up meeting to discuss suitability and other issues 	<ul style="list-style-type: none"> Pre-ketogenic diet: Weight and height assessed Baseline blood and urine collection One-month history of seizure activity Three-day diet history Introduction of vitamin/mineral supplement Review of medications and any necessary changes as carbohydrate content may interact with diet Prescription for equipment Dietary teaching and parental assignments completed 	<ul style="list-style-type: none"> MDT referrals: Referral to clinical psychology (if required) for both child and parents Assess need for home support. Liaise with neurology social worker. Liaise with public health nurse re equipment requirements. Play therapist input may be required during admission. 	<ul style="list-style-type: none"> Admission for commencement of ketogenic diet: Admitted for monitoring during commencement of diet. Nursing and dietary care plans to be completed by neurology CNS, ward staff nurses and dietitian. Education and training care plan for ketogenic diet Teaching plan for glucose/ketone monitoring on the ketogenic diet. 	<ul style="list-style-type: none"> Discharge and follow-up: Telephone contact/advice Once a month review at OPD – bloods required 3-6 monthly review at OPD with blood/urine work-up (refer to investigations of ketogenic diet) Monitor seizure activity

34.11.11 Appendix 11 2011 Paediatric Population and Paediatric Admissions

Geographical Area	Total Paediatric Population	Annual Admissions	(%)
Dublin (259,277) Kildare (54,252) Wicklow (32,972)	346,501	24,173 (includes tertiary admissions)	7%
Laos (20,978) Offaly (19,148)	40,126	2,464	6%
Longford (9,600) Westmeath (20,686)	30,286	2,887	9.5%
Cavan (18,242) Monaghan (14,326)	32,568	2,830	9%
Donegal (39,313)	39,313	4,074	10%
Sligo (13,907) Leitrim (7,316)	21,223	2,311	11%
South Tipperary	20,080	1,100	5.5%
Mayo (29,078)	29,078	2,867	10%
Galway (55,238) Roscommon (14,428)	69,666	6,025	9%
Louth (30,220) Meath (48,880)	79,100	3,780	5%
Kilkenny (22,513) Carlow (12,818)	35,331	2,356	6.6%
Limerick (41,417) North Tipperary (16,477) Clare (27,641)	85,535	5,238	6%
Kerry (31,222)	31,222	2,944	9.4%
Cork (115,879)	115,879	8,600	7.4%
Waterford (26,045)	26,045	3,548	13.6%
Wexford (35,014)	35,014	2,131	6%

Total paediatric population: 1,306,966