Paediatric Neurology Training Guide EPNS

Adapted from British Core training

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Contents

Core Training Programmes

Three generic components of paediatric neurology training have been identified as encompassing skills and knowledge that are relevant to all of the sub-speciality programmes within the paediatric neurology syllabus. Core programmes have been developed for neurophysiology, neuroradiology and neurogenetics.

There will be overlap with the requirements of the sub-speciality programmes and many of the core skills may be acquired within the context of one of the sub-speciality areas.

1. Neurophysiology Programme for Paediatric Neurology Trainees

Revision of Neuroanatomy

- Knowledge of the major subdivisions of the central and peripheral nervous systems

Basic Neurophysiology

- Basic knowledge of nerve conduction including ion channel function
- Synaptic function (inhibitory and excitatory) and the neuromuscular junction

Neuropharmacology

- Central nervous system neurotransmitters and drugs which modulate them
- Mode of action of drugs affecting the central and peripheral nervous systems

Neuroplasticity

- Basics of brain developmental and age dependant reorganisation processes

Standard EEG

- Indications and limitations of EEG as a diagnostic tool in a range of medical disorders
- Familiarity with EEG technology and equipment
- Neurophysiological basis of EEG signals and activation techniques
- Standard electrode nomenclature and standard montages used
- Know the normal pattern of EEG activity and sleep stages
- Recognize normal and non-epileptifom benign variants of EEG
- Artefacts in EEG recording
  Recognise EEG abnormalities (interictal and ictal) of focal and generalised epilepsies
- Role of EEG monitoring in PICU setting
  Neonatal EEG and evolution of maturational changes
- Write a factual report and give clinical conclusion in the end to referring physician with supervision

Video EEG telemetry and Ambulatory EEG

- Indications for long-term EEG monitoring and the limitations of these techniques
- Role of Video EEG in characterisation and classification of paroxysmal events
- Role of video EEG in pre-surgical evaluation of epilepsy
  Evaluate and interpret video recordings epileptic and non-epileptic seizures

Amplitude integrated EEG

- Indications and limitation on neonatal units
- Familiarity with technology and equipment
- Pattern recognition of normal aEEG and pathological findings

Clinical Neurophysiology for Epilepsy Surgery

- Role of scalp EEG in characterising and classifying seizures in pre-surgical assessment of epilepsy
- Limitations of scalp EEG in localising epileptogenic zone
- Strategies of multidisciplinary pre-surgical assessment for epilepsy surgery
- Be familiar with goals and risks of intracranial EEG monitoring (Subdural, Depth, Stereo EEG and Corticography), for precise localisation of seizure onset zone and functional mapping of eloquent cortex

Peripheral neurophysiology

- Physiology of nerve conduction, neuromuscular transmission and excitation -contraction mechanisms in muscle
- Clinical presentation and pathophysiology of diseases of the peripheral nerves, neuromuscular junction and muscles
- Anatomy of peripheral nerves and muscles with regard to electrode placement and needle insertion
- Techniques for study of peripheral nerves including sensory, motor, and F wave studies, H reflex, repetitive nerve stimulation
- Techniques of electromyography including recognition of neurogenic and myopathic disorders

Evoked potentials

Visual Evoked Potentials (VEP)

To understand the technical basis and methods of recording visual evoked potentials, appreciate when these tests may be used, and the expected changes from normal in a variety of pathological conditions

Electroretinogram (ERG)

To understand the physiological basis of the normal ERG, the technical aspects of its recording in children of all ages and its role in the investigation of neurological disorders.

Somatosensory Evoked Potentials (SSEP)
In spinal cord injuries: congenital, acute, for spinal monitoring in scoliosis surgery; prognostic functional assessments.

Transcranial Magnetic Stimulation (TMS)
Functional motor pathway assessments and therapeutic TMS) neuromodulation following perinatal and acquired stroke; possible emergent therapeutic uses in epilepsy; depression and anorexia.

Polysomnography and Multiple Sleep Latency Tests

- Classification and semiology of sleep disorders
- Normal EEG and polygraphic findings in sleep
- Indications for polysomnography and MSLT and the limitations of these techniques
- Stages sleep and recognition of features of common sleep disorders

Specific activities suggested to evidence completion of training in neurophysiology

Trainees are encouraged to participate in the following activities to support their training in neurophysiology:

Essential

- Regular attendance at EEG/neurophysiology MDT *
- Participation in EEG reporting sessions
- Completion of at least 2 case studies during training that focus on the neurophysiological aspects of diagnosis and management. (NB This could also form part of the training in other sub-speciality areas eg epilepsy or neuromuscular)
- Evidence of clinical presentation, audit or service evaluation where neurophysiology is the focus

Desirable

- Participation in neurophysiology teaching sessions when available
- Completion of British Distance Learning Unit 0 Introduction to Paediatric Neurology
- Virtual courses on EEG of ILAE
- Attending the EPNS teaching Course on Epilepsy and Neuromuscular problems

* Many units will have regular EEG meetings and trainees should endeavour to attend these when possible. In units where no regular on site meetings occur the trainee should be encouraged to visit of site meetings.

2. Neuroradiology Programme for Paediatric Neurology Trainees

By the end of their training, grid trainees will:

1. Understand the physical and technical principles behind commonly used imaging modalities - US, CT and MR, PET
2. Be familiar with the normal neuroanatomy of standard axial imaging (CT and MR) and sagittal and coronal MR
3. Be able to systematically describe CT and MR appearances
4. Be aware of common anatomical and developmental variants e.g. appearance of perivascular spaces, developmental venous anomalies
5. Know the normal myelination timetable as demonstrated on T2 and T1 MR
6. Recognise the normal development of the cortex from fetal to post-natal life

CT imaging

- **Be aware of the strengths and limitation of CT and with the CT appearances of the following:**

1. Cerebral oedema
2. Central and uncal herniation
3. Diffuse severe hypoxic - ischaemic injury - pattern reversal
4. Hydrocephalus - acute and chronic
5. Epidural haemorrhage
6. Sub-dural haemorrhage and appearances of inflicted brain injury
7. Intracranial infection
    a. Abscess
    b. Empyema
    c. Bacterial meningitis
    d. Herpes encephalitis
8. Tumours/SOL
9. Dural sinus thrombosis

MR imaging

- **Be familiar with commonly used sequences - T1, T2, FLAIR, GRE, SWI, DWI and MRS.**
- **Be familiar with the appearances of the following :**

1. Malformations
    a. Cortical malformations
        i. Lissencephaly (including cobblestone)
        ii. Polymicrogyria
        iii. Hemimegalencephaly
        iv. Focal cortical dysplasia
    b. Brainstem malformations
    c. Cerebellar malformations
    d. Chiari 1 and 2

    e. Neural tube defects
2. Neurocutaneous disorders
    a. TSC
    b. NF1
    c. Sturge Weber Syndrome
3. White matter disorders
    a. PVL
    b. MS/MOGAD/NMOSD and other immune related demyelinating diseases
    c. Understand the concept of hypomyelination vs delayed myelination
    d. Recognise classical MR phenotypes eg.
        i. X-ALD
        ii. MLD
        iii. VWM
        iv. PMD
        v. Mitochondrial leukodystrophies
4. Grey matter disorders
    a. Mitochondrial eg
        i. MELAS
        ii. POLG
        iii. Leighs
    b. Neuronal ceroid lipofuscinoses
    c. NBIAs
5. Stroke
    a. Arterial ischaemic stroke
    b. Moya Moya
    c. Carotid dissection
    d. Sagittal sinus thrombosis
    e. Cavernoma
    f. Normal MRA and MRV appearances
6. Brain tumours
    a. Medulloblastoma/ependymoma
    b. Pilocytic astrocytoma
    c. Glioblastoma
    d. DIPG
    e. Craniopharyngeoma
    f. DNET
    g. Optic pathway glioma
7. Spinal disorders
    a. LETM
    b. Intramedullary tumours
    c. Extra medullary tumours

    d. Syrinx
8. Brain damage patterns
    a. Neonatal HIE
    b. PVL
    c. Porencephaly
    d. Multicystic encphalomalacia
    e. Hydranencephaly
    f. Hippocampal sclerosis
9. CNS infection
    a. Congenital infection esp. CMV,HSV, toxoplasmosis
    b. Herpes simplex encephalitis
    c. Abscess
    d. Meningitis

- **Understand the role of Advanced Imaging techniques in the assessment and Management of Children with Neurological Disorders:**
    - 3T MR
    - fMRI
    - PET/SPECT
- **Be familiar with evolving MR modalities eg DTI, FA, MTI**

Specific activities suggested to evidence completion of training in neuroradiology

Trainees are encouraged to participate in the following activities to support their training in neuroradiology:

Essential:

- Regular attendance at neuroradiology MDT *
- Attendance at neuro-oncology MDT
- Completion of at least 2 reflective case notes during training that focus on radiological aspects of diagnosis and management
- Evidence of clinical presentation, audit or service evaluation where neuroimaging is the focus

Desirable

- Attendance at neuroradiology teaching sessions
- Completion of British Distance Learning Unit 0 Introduction to Paediatric Neurology Attendance of EPNS Webinars : “Migration problems and it’s genetics” and “Brain malformations and it’s Genetics”

* Many units will have weekly neuroradiology MDTs and trainees should endeavour to attend these when possible. In units where no regular on site meetings occur the trainee should be encouraged to visit of site meetings.

3. Core Genetics Programme for Paediatric Neurology Trainees

At the end of their training, a trainee should understand and be confident in discussing:

- Basic patterns of inheritance – AR, AD, XL, mt
    - Concepts of variable expression, non-penetrance and age-related penetrance
    - Clarification of the term 'sporadic'
    - New dominant mutations
    - Significance of consanguinity
    - Gonadal / somatic mosaicism
    - Trinucleotide repeat disorders
- Consent to genetic testing
- Testing landscape: – era of rapid change
    - Databases of sequence variation such as GnoMAD or EXAC that give information on whether the variant occurs in the "normal " population
    - Lack of predictive value of Single nucleotide polymorphisms for 'personalised medicine'
    - Methods of prenatal diagnosis: including pre-implantation genetic diagnosis
- Genetic sequencing
    - Sanger sequencing
    - Exome sequencing
    - Whole genome sequencing – and the importance of the trio (child, mother and father)
    - Panel sequencing
- Interpretation and further elucidation of sequencing results:
    - Incidental findings
    - Variants of unknown significance
- Interpretation of sequence variants on the basis of:
    - Familial segregation / rarity / ethnicity / in silico prediction / previous association with disease / animal models / functional assays
    - Possibility of digenic inheritance
- Expansion of phenotypes (with Next Generation Sequencing) and impact disease classification by phenotype or by genotype or pathway
- Gene / protein networks
- Increasing possibility of treatments
- Giving the information to families to allow informed consent for genomic investigations. The principles of this can be applied to other investigations such as MRI and metabolic. With particular reference to discussing the limits of investigation, false positive/negatives.

Key activities

Trainees should be encouraged to participate in the following acitivites to support their training in neonatal neurology:

Essential

- Reflective case log of 5 patients with an underlying genetic problem
- 2 case based discussions that cover genetic problems
- Attend genetic boards/rounds for case discussion
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Desirable

- Attend a genetic counselling session
- Completion of British Distance Learning Unit Genetics

4. Developmental Neurology – the normal, the abnormal and learning disorders

1. Undertake a detailed history and assessment of a child /adolescent with a suspected developmental problem, assessing motor and cognitive skills using established tools. Being able to make an interpretation of developmental and neuropsychological tests.

2. Understanding the function of an MDT and role of each person (e.g. geneticist, psychologist, physio- speech and occupational therapists) and the role of the PN in it. Use relevant referral pathways and be aware of rapid changes of systems.

2. Assess, investigate and diagnose a child presenting with more common developmental problems including global developmental delay, language delay, specific learning disorders and being able to assist an MDT to make a specific diagnosis and care for management in disorders such as autism, fragile X, ADHD, trisomy 21.
3. Differentiate delay from regression and being able to initiate the diagnostic work up for both routes.

4. Evaluate, initiate and interpret investigations including neuroimaging, genetic testing, neurophysiology and metabolic. Discuss the limitation in the diagnosis of developmental problems.

5. Discuss management and prognosis of more common developmental conditions as above. Be aware of newer treatment options for certain problems e.g. genomic treatment for Angelman.

6. Understand the multisystem nature of developmental disorders (eg. hypothyroidism in Downs Syndrome), know how to assess and monitor.

7. Manage evolving phenotypes of developmental problems throughout infancy to adulthood.

7. Assess and manage a child with secondary psychiatric or behavioural problems and know which members of MDT should be involved for help / how to access them.

8. Assess and investigate a child presenting with abnormal head growth (as micro-macrocephaly, plagiocephaly, craniosynostosis) be aware of differential diagnosis and how to evaluate.

Key activities

- Case log reflecting the spectrum of developmental problems in out patients clinics as well as in patient care
- Attendance at specialist developmental clinics within an MDT
- Completion of assessments during training that cover diagnosis and management

Desirable

- Completion of on line course EPNS/McKeith: Principles of Child neurology in infancy
- Case report or presentation of a child with developmental problems
- Course for acquiring capacity to do developmental testing (as Bayley tests etc)
- Documentation of the investigation of intellectual disability.
- Attend EPNS Webinars “ADHD” and “Autism”

Many neurological disorders overlap with the above (learning disorders in tuberous sclerosis, epilepsy etc)

5. Neonatal and Fetal Neurology Programme for a Grid Trainee in Paediatric Neurology

At the end of their training a trainee in paediatric neurology should:

1. Be confident in the neurological examination of the preterm and term infant.

2. Understand the mechanisms, patterns, timings and range of outcome for acute profound and chronic partial hypoxic ischaemic brain injury.

3. Formulate a differential diagnosis and directed investigative plan for neonatal encephalopathy.

4. Know the ILAE classification for neonatal seizures, and the abnormal movements that are not likely to be seizures.

5. Draw up a differential diagnosis and directed investigative plan for neonatal seizures.

6. Be able to interpret investigations and develop a management plan for neonatal epilepsy syndromes.

7. Understand the nature of arterial ischaemic stroke (AIS) and cerebral venous sinus thrombosis (CVST) presenting in neonates. Be able to make a diagnostic plan and interpret investigations and formulate a management plan.

8. Be able to assess both the floppy and stiff baby. Be familiar with the limitations of investigations and be able to formulate a strategy for management.

9. Understand the pathogenesis of neural tube defects; be confident in the examination of affected infants and familiar with outcomes and key aspects of their management and co-morbidities.

10. Be familiar with patterns of brain malformation occurring at different stages of development, their aetiology, co-morbidities and outcomes.

Acute neurology

- Be familiar with the challenges in the management of status epilepticus in newborn infants at different gestational ages, including patterns of EEG, nature of MR abnormalities and therapeutic choices
- Have attended MDT meetings to discuss prognosis and possible withdrawal/reorientation of care in infants with severe neurological disorders including those with possible CNS malformation, HIE or neuromuscular disorders
- Have experience in counseling/discussion with parents regarding long term prognosis of neurological disorders presenting in neonates, uncertainty in prognosis and range of outcomes

Transition and adult neurology

- Have experience of maternal imaging of foetal brain with MR / ultrasound, and antenatal genomic investigation
- Be aware of the role of the foetal MDT, the referral pathways and strategies that exist to support parents with antenatally diagnosed neurological disorders

Neuropsychological and neuropsychiatric aspects

- Be aware of the cognitive and behavioural consequences of pre-term /severely dysmature birth
- Be aware of associated cognitive and behavioural effects following 'insults' to the developing brain

Neurodisability

- Be familiar with the neurodevelopmental sequelae of prematurity
- Know the risk factors for the development of cerebral palsy
- Recognize patterns of brain injury that occur in the neonatal period and their neurodevelopmental consequences
- Have experience of post discharge management of an infant with myelomeningocele, including management of bladder and bowel
- Be aware of the neurodevelopmental consequences of hydrocephalus and the important elements of management

Key activities

Trainees should be encouraged to participate in the following acitivites to support their training in neonatal neurology:

Essential

- Reflective case log of 10 neonatal neurology and 3 fetal neurology patients
- 2 case based discussions that cover neonatal neurological problems
- Attend a neonatal neurology or development follow-up clinic
- Attend at least 3 radiology meeting in which neonatal neuroimaging is reviewed
- Attendance at neonatal course (e.g. teaching course EPNS, BPNA NeoNATE course)
- Attend EEG meetings (3) in which neonatal EEG is reviewed

Desirable

- Attend a fetal / antenatal counselling session
- Completion of British Distance Learning Unit 2 Neonatal Neurology
- Completion of EPNS/McKeith on line course of Child Neurology in Infancy
- Attend EPNS Webinar on neonatal Neurology

6. Epilepsy Programme for a Trainee in Paediatric Neurology

At the end of their training a trainee in paediatric neurology should:

1. Be able to undertake a detailed history and assessment of a child presenting with paroxysmal events.

2. Be aware of the wide differential diagnosis of paroxysmal events in childhood, know the issues surrounding misdiagnosis in epilepsy and understand the effective management of diagnostic uncertainty and risk.

3. Be familiar with ILAE classification of epileptic seizures and of the epilepsies.

4. Be aware of age related epilepsy syndromes (neonatal, infantile, childhood and teenage) and be able to make epilepsy syndrome diagnoses where possible.

5. Be aware of the wide range of aetiologies underlying the epilepsies and initiate appropriate investigations for children presenting with epilepsy at all ages.

6. Be familiar with normal EEG patterns throughout the paediatric age range and have a knowledge of abnormalities seen on EEG. Be able to interpret an EEG report in the context of clinical information.

7. Be familiar with the co-morbidities of the epilepsies and aware of the importance of assessment.

8. Be able to choose and manage appropriate antiepileptic medication.

9. Be familiar with management of drug resistant epilepsies and referral pathways to specialist epilepsy clinics.

10. Have knowledge of epilepsy surgery referral criteria and have experience of management of children within an epilepsy surgery programme.

11. Understand the role of dietary treatment of the epilepsies, and criteria for referral. Be familiar with the management of children on the ketogenic diet for epilepsy, more specifically in case of acute illness or in perioperative situations.

12. Be familiar with role of vagal nerve stimulator in management of childhood epilepsies.

Acute neurology

- Be able to manage acute convulsive seizures and status epilepticus
- Be able diagnose and manage non-convulsive status epilepticus
- Understand how to diagnose, monitor and treat refractory status epilepticus and status epilepticus in ICU patients
- Know the systemic complications of status epilepticus
- Recognise the differential diagnosis of status epilepticus
- Be aware of disorders that may present with explosive onset epilepsy eg POLG , FIRES and related disorders
- Understand the indications, limitations and complications of the use of rescue medication for seizures and be familiar with the specific medications that may be used

Transition and adult neurology

- Be aware of issues in managing teenagers with epilepsy and have knowledge and experience of transition of care of epilepsy to adult services
- Be aware of epilepsies that more commonly present in adult life
- Know the prognosis and natural history of epilepsies that continue into adult life
- Understand the management challenges for young adults with severe epilepsies transitioning into adult services eg Lennox Gastaut /Dravet syndromes

Neuropsychological and neuropsychiatric aspects

- Be familiar with and able to recognise the common psychological and psychiatric consequences of a diagnosis of epilepsy
- Have experience of joint working neuropsychology in the management of children with epilepsy
- Have experience of management of a child/young person with severe behavioural difficulties and epilepsy
- Know how to diagnose, evaluate and manage a child with a cognitive epilepsy such as LKS / ESES eg joint working with neuropsychology, SALT and education in the management
- Be able to diagnose and manage non-epileptic attack disorder particularly in patients with co-existing epilepsy

Neurodisability

- Be confident in the assessment and management of a child with seizures and underlying neurodisability (Epilepsy Plus)
- Be familiar with the long-term developmental consequences of symptomatic epilepsy syndromes and their management

- Recognise the impact of 'benign' epileptic disorders on learning and behaviour and be able to work with colleagues in other agencies to formulate strategies for support and management
- Be aware of the potential neurodevelopmental sequelae of epilepsy surgery

Key activities

Trainees should be encouraged to participate in the following activities to support their training in childhood epilepsies:

Essential

- Reflective case log of 50 patients with a range of epilepsy syndromes presenting both acutely to inpatient settings and to outpatients with a mix of secondary and tertiary patients
- Attendance at epilepsy specialist clinics including epilepsy surgery, ketogenic diet, teenage epilepsy and transition clinics
- Attendance and participation in national or regional epilepsy network meetings
- Attending of the EPNS teaching course on Epilepsy or a similar national or international course (for example ILAE courses)

Desirable

- Evidence of audit or service evaluation that relates to children with epilepsy
- Completion of Paediatric Epilepsy Training 1, 2 and 3
- Completion of British Distance Learning Unit 6 Epilepsy
- Completion of 'Expert to Expert' Epilepsy course
- Attend EPNS Webinar

7. Movement Disorder Programme for a Trainee in Paediatric Neurology

At the end of their training a trainee in paediatric neurology should:

1. Understand the relevant neuroanatomy and neurophysiology in relation to aetiology and treatment of movement disorders (MD).

2. Be able to undertake a detailed history and assessment of a child with a suspected MD.

3. Be able to assess functional motor skills and outcomes with established tools.

4. Be able to recognise and evaluate normal variants in motor development, and be confident in interpretation of 'abnormal' results in otherwise healthy children to ensure children are not submitted to unnecessary investigations.

5. Be able to assess, investigate and diagnose a child presenting with common MD phenotypes including tics, stereotypies, dystonia, athetosis, chorea, ataxia, myoclonus and spasticity.

6. Be able to interpret CSF neurotransmitter results.

7. Be familiar with the management of common movement disorders including orthoses and therapies, medication, botulinum neurotoxin and functional neurosurgery (including deep brain stimulation and intra-thecal Baclofen).

8. Recognise the natural history of MDs from foetal to adult life.

<u>Acute neurology</u>

- Be able to recognize and manage status dystonicus
- Know the common causes of acute dystonic reactions to medication
- Be able to recognize, investigate and manage acute onset movement disorders e.g. chorea

<u>Transition and adult neurology</u>

- Be able to assess, investigate and diagnose early onset presentations of common adult disorders e.g. Huntingtons Disease, hereditary ataxias,
- Be familiar with the current management of Parkinson's disease

Neuropsychological and neuropsychiatric aspects

- Be aware of progressive disorders that may present with neuropsychiatric features before the onset of MD
- Be aware of the neuropsychiatric features associated with MD
- Be familiar with current neuropsychiatric and psychological approaches to the management of tic disorders and Tourette syndrome
- Understand the role, and potential complications and drug interactions of psychotropic drug treatment for neuropsychiatric co-morbidities of MDs

Neurodisability

- Be familiar with the long-term consequences of different movement disorders on learning, behaviour and performance
- Be able to co-ordinate multidisciplinary management in a child with a movement disorder that affects their cognition, motor and social skills and impacts on their participation
- Be familiar with the management of involuntary movements and hypertonia in children with underlying developmental disorders, ie cerebral palsy, and how their presentation may evolve with time
- Be confident in distinguishing neurodevelopmental disorders that may present with abnormal movements, ie autistic spectrum disorders from new onset neurological disorders
- Be familiar with progressive neurological disorders whose juvenile forms may have subacute presentations that may be misdiagnosed as developmental disorders, ie Juvenile Huntingdon's disease

Key activities

Trainees would be expected to produce the following evidence to support their training in movement disorders:

Essential

- Reflective case log of 20 patients with MD presenting acutely or as out-patients

Desirable

- Attendance at specialist MD clinics
- Evidence of audit or service evaluation that relates to children with MD
- Attendance at a Paediatric Movement Disorder special interest group
- Completion of British Distance Learning Unit 4 Central Motor Deficits
- Attendance of teaching course EPNS on metabolic diseases
- Completion of EPNS/McKeith on line course of Child Neurology in Infancy
- Attend EPNS Webinar

8. Neuromuscular Programme for a Trainee in Paediatric Neurology

At the end of their training a trainee in Paediatric neurology should:

1. Be able to undertake a detailed history and assessment of a child with a suspected neuromuscular disorder (NMD). This will include assessment of functional motor skills with established tools, evaluation of muscle power and joint range and recognition of different patterns of muscle involvement.

2. Be able to recognize and evaluate normal variants in motor development, and be confident in interpretation of 'abnormal' results in otherwise healthy children to ensure children are not submitted to unnecessary investigations.

3. Be able to assess, investigate and diagnose a child presenting with more common NMD including dystrophin related muscular dystrophy, Spinal Muscular Atrophy (SMA), inherited peripheral neuropathy (CMT), Myasthenia Gravis and congenital myotonic dystrophy.

4. Be able to evaluate, initiate and interpret investigations including neurophysiology and muscle biopsy reports and muscle imaging studies in a child with possible myopathy or neuropathy.

5. Be able to discuss management strategies and prognosis of more common neuromuscular conditions (as outlined in 3. above).

6. Recognize and evaluate the systemic, metabolic and mitochondrial disorders that can affect neuromuscular function.

7. Understand the multisystem nature of NMD and have knowledge of how to assess and monitor potential complications including cognitive, respiratory, cardiac and postural abnormalities and the strategies for their management.

8. Understand the use and limitation of genetic studies for diagnosis and management of children with NMD. This will include antenatal screening, carrier testing and the approach to presymptomatic individuals.

9. Understand, and be familiar with, referral pathways and multidisciplinary networks for the management of children with NMD, including national networks for audit and research, international standards of care in DMD, SMA etc, patient registries, relevant charities and NCG centres.

<u>Acute neurology</u>

- Be able to assess and initiate investigation and management in a child presenting with acute paralysis or rhabdomyolysis
- Be able to assess and initiate management for the acute systemic complications of

neuromuscular disease including acute respiratory failure, cardiomyopathy, fracture and fat embolus syndrome
- Be familiar with the role of critical care and palliative care teams in management of acute complications of NMD and in drafting acute and long-term care plans for individual neuromuscular patients
- Recognize the symptoms and signs of critical illness polyneuropathy and myopathy

Transition and adult neurology

- Understand the needs of the patient and family during the transition process and have experience of having managed one or more patient with NMD through handover to adult care/services
- Be familiar with the evolution of childhood onset disorders in young adults ie GI disturbance in DMD, myotonia in congenital myotonic dystrophy
- Be aware of the "adult" onset NM disorders that may present in childhood

Neuropsychological and neuropsychiatric aspects

- Know the associated cognitive and behavioural profiles of specific NMD, in particular DMD and congenital myotonic dystrophy.
- Recognise the common neuropsychiatric co-morbidities in NMDs
- Understand the role of neuropsychology and /or psychiatry in the management of children with NDDs and their families

Neurodisability

- Be aware of the shared pathways for children with NMD and other ND. Be comfortable working with both specialist and community multidisciplinary teams, recognising their important roles in supporting children and families with NMD
- Be confident in highlighting different approaches in managing certain elements of care ie postural support, contracture management, use of orthotics and orthopaedic intervention in children with NM disorders in comparison to other conditions associated with ND ie cerebral palsy
- Be aware of the important role that palliative care teams play in supporting children with NMD

Key activities

Trainees should be encouraged to participate in the following activities to support their training in neuromuscular disorders:

Essential

- Brief case log of 20 patients with both acquired and inherited NMD presenting acutely and in outpatients
- Attendance at 8 specialist NM clinics, including at least 2 adult clinics
- Attendance at a national or regional muscle special interest group meeting

Desirable

- Completion of British Distance Learning Unit 5 Neuromuscular Disorders
- Completion of EPNS Teaching course on neuromuscular diseases
- Audit or service evaluation that relates to children with NMD
- Case report or presentation of child with NMD
- Attend EPNS Webinar

It is also noted that a number of aspects overlap with generic management of children with complex neurodisability / other neurodegenerative diseases and so learning form other subspecialities may address important elements of multidisciplinary working, management of disability, end of life planning and could be cross referenced for NMD competencies.

9. Neuroinflammation Programme for a Trainee in Paediatric Neurology

At the end of their training a trainee in paediatric neurology should:

1. Be competent in examining the child or infant with acute and chronic neuroinflammatory conditions.

2. Be able to establish correct diagnosis by clinical assessment, and basic investigations for acute syndromes:
   - Autoimmune encephalitis and the range of immune-mediated CNS syndromes and associated antibodies
   - The range of acquired demyelination syndromes to include optic neuritis, transverse myelitis, neuromyelitis optica, clinically isolated syndromes and multiple sclerosis and their imaging characteristics
   - Acute neurological syndromes with systemic inflammatory disorders

3. Be able to initiate acute and maintenance immunotherapy and required monitoring.

4. Be aware of current concepts in CNS inflammation.

<u>Acute neurology</u>

- Be familiar with acute presentations that may be the result of a CNS inflammatory disorder. For example:
  - NMDAR encephalopathy
  - Limbic encephalitis
  - Transverse myelitis
  - Optic neuritis
- Be aware of the indications for and complications of immunotherapies that may be required in CNS inflammatory disorders e.g. Steroids and rituximab

<u>Transition and adult neurology</u>

- Be aware of the more common teenage presenting CNS inflammatory disorders
- Be familiar with current approaches to diagnosis and management of MS in adults
- Understand the needs of the patient and family during the transition process and have experience of having managed one or more patient through this

<u>Neuropsychological and neuropsychiatric aspects</u>

- Be aware of the CNS inflammatory disorders that may present with neuropsychiatric features
- Recognise and be able to evaluate and manage the psychological and neuropsychiatric aspects of CNS inflammatory disorders and their treatment

- Have experience of joint working with psychology and/or psychiatry in the management of patients with CNS inflammation

Neurodisability

- Be aware of the impact of neuro-inflammatory disorders on learning, behaviour and day to day activities
- Be able to develop management strategies that reflect the varying symptoms and signs that may impact on performance and inclusion
- Be able to recognize the features of a neuro-inflammatory disorders in a child with pre-existing developmental difficulties and be able to co-ordinate appropriate investigations and treatment

Key activities

Trainees should be encouraged to participate in the following activitiess to support their training in neuroinflammation:

Essential

- Reflective log of 5 cases of autoimmune encephalitis
- Reflective log of 3 cases of systemic immune mediated disorders with a CNS presentation
- Reflective log of 10 cases of acquired demyelination syndromes including MS
- Attend a national or regional Childhood Inflammation Disorders special interest group meeting

Desirable

- Involvement in audit or quality improvement project or protocol development
- Completion of British Distance Learning Unit 8 Inflammation & Infection of the CNS or EPNS immunology course
- Attendance of EPNS teaching course in Neuroimmunology
- Attendance at Chronic inflammation MDT e.g. MS (including imaging)
- Attend EPNS webinar on OMS

If a trainee is unable to access some aspects of training, they may need to arrange a clinical placement in a unit with a higher case load to access the range of experience required

10. Neurodegenerative and Neurometabolic Diseases Programme for a Trainee in Paediatric Neurology

At the end of their training a trainee in Paediatric Neurology should:

1. Be familiar with the terminology used in the classification of neurodegenerative diseases (NDD) and neurometabolic disorders.

2. Be able to undertake a detailed history and assessment of a child with suspected NDD presenting at any age from the neonatal period through to young adulthood.

3. Be able to distinguish “pseudo-regression” from true regression and know how to use investigations to enable this distinction (eg non-convulsive status, hydrocephalus, brain tumour).

4. Understand how to investigate suspected neurometabolic disease and be familiar with joint working with metabolic disease specialists.

5. Be familiar with the neuroimaging features of the main categories of NDD eg leukodystrophy, neurodegeneration with brain iron accumulation (NBIA), NCLs, mitochondrial disease or peroxisomal disorders.

6. Be able to use and interpret investigations for NDD.

7. Recognize the important treatable NDDs and Neurometabolic disorders and understand how to investigate these in a timely manner.

8. Have some experience of symptomatic management of NDDs.

9. Be aware of experimental or early clinical trials for rare NDDs.

10. Be aware of relevant patient support groups for childhood presenting NDDs.

Acute neurology

- Be familiar with the diagnosis and management of neurodegenerative or neurometabolic diseases which may present acutely.
    a. Status epilepticus in POLG and other metabolic disorders
    b. Acute encephalopathy in mitochondrial, urea cycle, amino/organic acid disorder and fatty acid oxidation disorders
    c. Acute paralysis +/- rhabdomyolysis in metabolic disorders
    d. Multi system failure in metabolic disorders
    e. Status dystonicus

- Be aware of the complications of neurodegenerative diseases that may result in acute presentation

Transition and adult neurology

- Be aware of NDDs where survival into adult life is likely eg CLN3, Freidreichs ataxia, AT
- Understand the needs of the patient and family during the transition process and have experience of having managed one or more patient through this
- Be aware of the "adult" neurodegenerative disorders that may present in childhood eg HD, PD
- Be aware of NDDs that usually present in teenage or young adults eg Wilsons, HD, MERRF/MELAS

Neuropsychological and neuropsychiatric aspects

- Know the NDDs that may present with cognitive/behavioral/psychiatric features before other neurological features develop
- Understand how to evaluate suspected cognitive decline
- Recognise the common neuropsychiatric co-morbidities in NDDs
- Understand the role of neuropsychology and /or psychiatry in the management of children with NDDs and their families

Neurodisability

- Be able to recognize neurometabolic and ND disease in a child with pre-existing learning difficulties or disability
- Recognize when a child with what was thought to be a static developmental disorder progresses and requires further assessment/investigation
- Be confident in contributing to MD meetings and assessments for children with NDD with associated learning, developmental and behavioural difficulties

Key activities

Essential

- Reflective case log of 6 patients with confirmed NDD
- Reflective log of 4 patients with “pseudo regression” (See 3 above)
- Attendance at least 1 x neurogenetics club or special interest group of a regional/national MDT
- Attendance of at least 2 specialist metabolic diseases clinic (could be an adult clinic if no paediatric clinic accessible))

Desirable

- Evidence of audit or service evaluation that relates to children with NDD
- Case report or presentation of NDD/metabolic patient
- Completion of British Distance Learning Unit 9 Metabolic, Nutritional and Systemic Disease
- Attendance of EPNS course on neurometabolic disease
- Attend EPNS webinar

11. Neurovascular Disorders Programme for a Trainee in Paediatric Neurology

At the end of their training a trainee in paediatric neurology should:

1. Have developed an understanding of epidemiology, presentation, etiology, investigation and management of neurovascular conditions in childhood

2. Know the risk factors for arterial ischaemic stroke (AIS), cerebral venous thrombosis (CVST) and other occlusive arteriopathies including spinal stroke.

3. Be familiar with imaging characteristics, classification and investigation of AIS, CVST and other arteriopathies and their differential diagnoses.

4. Be aware of therapeutic strategies for management of both paediatric and adult stroke.

5. Understand the key differences between perinatal stroke and stroke in older infants and children.

6. Understand the causes and presentation of nontraumatic intracranial haemorrhage, including AV malformation and aneurysms.

7. Know the principles of neurorehabilitation, investigation and management of traumatic hemorrhage (see ABI/rehab training programm).

8. Be aware of the principles in management of intracranial and intraspinal AV shunts (particularly high flow AV shunts such as Vein of Galen malformation).

<u>Acute neurology</u>

- Be confident in the initial assessment and management of a child presenting with acute stroke
- Be aware of hyper acute investigation and management of stroke including role of thrombolysis and mechanical thrombectomy
- Be aware of the strategies for managing acute non traumatic haemorrhage in critical care setting
- Be aware of referral pathways for management of severe AV malformation that present acutely e.g. Vein of Galen malformations

Transition and adult neurology

- Be familiar with management, especially hyperacute management, of adult stroke
- Understand the needs of a young adult with arteriopathy during the transition process and have experience of having managed at least one patient through handover to adult care/services
- Be aware of the "adult" risk factors for stroke/arteriopathy that may present in childhood e.g. SLE other vasculitides

Neuropsychological and neuropsychiatric aspects

- Be aware of the cognitive and behavioural consequences of acute childhood stroke
- Be aware of more insidious cognitive and behavioural effects of arteriopathies
- Understand the role of neuropsychological assessment in monitoring disease progression and treatment

Neurodisability

- Be able to recognize neurovascular disorders with pre-existing learning difficulties or disability especially where presentation may be subacute and associated with gradual decline e.g. Moya Moya syndrome
- Be familiar with developmental disorders that have a higher risk of neurovascular complications e.g. Down Syndrome
- Be able to tailor appropriate diagnostic investigations in a child with neurodisability and suspected stroke or venous sinus thrombosis
- Be able to lead multi-disciplinary meetings to support discharge planning, goal setting and long-term management in children with stroke

Key activities

Trainees will need to be expected to produce the following evidence to support their training in neurovascular disorders*:

Essential

- Case log of 20 patients (including 6 adult cases) with a wide range of neurovascular disorders
- Attend 2-3 Neurovascular MDT meetings with diagnostic and interventional radiologists and neurosurgeons, presenting cases and implementing agreed action plans
- Attendance of a national or regional cerebrovascular special interest group meeting

Desirable

- Visit to supraregional centre to observe mechanical thrombectomy and embolization procedures
- Evidence of audit or service evaluation that relates to children with neurovascular disorders
- Case report of child with neurovascular disorder
- Complete Distance Learning Unit 7 Cerebrovascular disease, trauma and coma
- Attend the EPNS teaching course Acute Neurology
- Attend EPNS webinar “Ischaemic stroke” and Haemorrhagic stroke- vascular malformations”

If a trainee is unable to access some aspects of training, they may need to arrange a clinical placement in a unit with a comprehensive paediatric neurovascular practice

12. Neuro-oncology Programme for a Trainee in Paediatric Neurology

At the end of their training a trainee in paediatric neurology should:

1. Know the common clinical presentations of CNS tumours in children and young adults.

2. Understand the reasons why delayed diagnosis of CNS tumours occurs.

3. Be familiar with the emergency management of acutely presenting CNS tumours.

4. Appreciate the role of multidisciplinary pre-surgical evaluation by the neuro-oncology MDT.

5. Be able to identify classical CNS tumour appearances on neuro-imaging and appreciate the role of advanced imaging modalities (see neuroradiology curriculum).

6. Be aware of the differential diagnosis of disorders that may initially be suspected to be a CNS tumour (including patients presenting with epilepsy).

7. Be familiar with perioperative management of CNS tumours and common complications.

8. Be able to diagnose and manage Cerebellar Mutism Syndrome

9. Be aware of current adjunctive treatment modalities for brain tumours
    a. Photon radiotherapy
    b. Proton radiotherapy
    c. Stereotactic radiotherapy
    d. Chemotherapy

10. Be aware of the common late-effects of treatment for a CNS tumour including neuropsychiatric morbidities and learning disabilities.

11. Be aware of the role of molecular diagnostics in stratifying treatment for brain tumours.

12. Be familiar with the most prevalent genetic tumour predisposition disorders such as:
    a. NF1
    b. TS
    c. Li- Fraumeni
    d. Gorlin syndrome

    See also chapter on neurocutaneous disorders

13. Be aware of the approach to management for congenital brain tumours.

14. Understand the role of radiological surveillance and clinical monitoring in the management of brain tumours.

15. Be aware of gene therapy and other experimental treatments for brain tumours.

16. Have experience of end of life management for patients with brain tumours.

17. Understand the particular management issues for a young adult with a "paediatric brain tumour" eg medulloblastoma.

18. Recognize and be able to investigate and treat opsoclonus myoclonus syndrome

Acute neurology

- Be familiar with acute presentation of brain tumours with
    - Acute hydrocephalus
    - Intracerebral haemorrhage
    - Other signs of raised intra cranial pressure
    - Acute onset of Seizures or focal neurology

- Be aware of acute management strategies ie use of intraventricular shunts and drains and steroids
- Be aware of acute complications of surgery for children with brain tumours
    - Cerebral swelling and oedema
    - DI, SIADH and cerebral salt wasting
    - Cerebellar Mutism syndrome
    - Eye movement disorders

Transition and adult neurology

- Be familiar with the long-term consequences of chemotherapy and radiotherapy and their impact in the long-term, ie endocrine issues affecting growth and fertility
- Be aware of strategies for long-term follow-up of children with both benign and malignant brain tumours
- Understand the needs of the patient and family during the transition to adult services and have experience of having managed one or more adolescents through the transition process

Neuropsychological and neuropsychiatric aspects

- Be aware of the cognitive/behavioural/psychiatric features of brain tumours and their treatment
- Understand how to evaluate suspected cognitive decline following brain tumours
- Recognise the common neuropsychiatric co-morbidities of brain tumours and the role of neuropsychology and /or psychiatry in their management

Neurodisability

- Be able to recognize a brain tumour in a child with pre-existing learning difficulties or disability
- Be able to tailor appropriate diagnostic investigations in a child with neurodisability and suspected brain tumour
- Be able to contribute to discussions about long-term care and treatment goals, and to assist families in assessing the risk and benefits of treatment

Key activities

Trainees should be encouraged to participate in the following activities to support their training in neuro-oncology:

Essential

- Attendance at 5 neuro-oncology MDT meetings
- Reflective case-log of 10 patients with a brain tumour including at least one of the following: medulloblastoma, optic pathway glioma, craniopharyngioma, posterior fossa pilocytic astrocytoma, diffuse pontine glioma, non-tumour SOL

Desirable

- Attendance at a neuro-oncology follow up clinic
- Evidence of audit or service evaluation that relates to children with a brain tumour
- Completion of British Distance Learning Unit 11 Neuro-oncology
- Attendance of EPNS Webinar “CNS tumors”

13. Non neuro-oncological neurosurgical program in Paediatric Neurology

At the end of their training a trainee in paediatric neurology should:

1. Recognize common spinal problems such as dysraphia, spinal tumors, spinal infarctions, know how to investigate and understand treatments especially know about neurosurgical indications

2. Be able to evaluate a child with back pain

3. Assessing the child with hydrocephalus, being able to differentiate aetiologies. Differentiation of shunt complications (especially blockage from other disorders)

4, Recognize complications after shunt placements on long term

5, Supporting the neurosurgeon in multidisciplinary management.

Key activities

- Reflective case log of at least 5 children of different ages and aetiologies with hydrocephalus and shunt
- Case log of children spinal problems of any kind (such as tumors, dysraphism, vascular problems)

Desirables
- Attend team meetings including children with intracranial pressure/disorders
  - Attend MMC clinics
-Attend EPNS webinars “Spinal problems” and “Hydrocephalus”

14. Neurocutaneous program for a Trainee in Paediatric Neurology

At the end of their training a trainee in paediatric neurology should:

1. Undertake a detailed history and assessment of a child /adolescent with a suspected neurocutaneous disorder (NCD).
2. Be aware of the multiorgan problems in the most frequent ones (e.g. neurofibromatosis, tuberous sclerosis and Sturge Weber (SWS). Be aware of neurocutaneous disorders which predispose to CNS tumor (e.g., Gorlin syndrome, Cowden syndrome, neurocutaneous melanosis, v Hippel Lindau disease

3. Be aware of the multidisciplinary management of these disorders and the leading role of the PN in it.
4. Evaluate, initiate and interpret investigations including neuroimaging, genetic testing, neurophysiology and neuropsychology.

5. Discuss management and prognosis of the above-mentioned most common NCD. Be able to outline an appropriate surveillance and be aware of newer treatments for certain problems e.g. everolimus in TSC, MEK inhibitors in NF)

5. Manage evolving phenotypes of neurocutaneous problems throughout infancy to adulthood
6. Be aware of genomic investigations for both Mendelian inheritance e.g. TSC and also Mosaic disorders such as Sturge-Weber syndrome

Key activities

- Case log reflecting the spectrum of neurocutaneous disorders in out patients clinics as well as in patient care
- Reflective case log of at least 2 children each with neurofibromatosis, tuberous sclerosis and Sturge Weber
- Attendance at specialist rounds to discuss neuroimaging findings, kidney problems etc
- Completion of assessments during training that cover diagnosis, management and follow up
- Attendance at a follow up clinic for neurocutaneous disorders especially NF, TS and Sturge Weber
- Recording of the surveillance required for NCD including TSC, NF1 and SWS

Desirable

- Visiting the EPNS webinar on neurocutaneous disorders
- Case report or presentation of a child with neurocutaneous problem
- Attend EPNHS webinar on “Neurofibromatosis” and “Tuberous Sclerosis”

15. Neuro-ophthalmology program for a Trainee in Paediatric Neurology

At the end of their training a trainee in paediatric neurology should be able to:

1. Take a detailed history and perform a clinical neuro-ophthalmological (NO) examination (e.g. fundoscopy, oculomotor movements, visual fields, examination with Frenzel , RAPD) in a child with NO problems (not an expert assessment but that which a PN performs).
2. Be able to recognize pupillary anomalies, ptosis, iris and conjunctival problems
3. Be able to differentiate oculomotor problems e.g. strabismus, cranial nerve deficits, supranuclear problems)
4. Recognize developmental problems as Duane syndrome, Marcus Gunn jaw winking, physiological anisocoria
5. Be able to recognize retinal and optic nerve problems in fundoscopy – especially papilledema
6. Understand the role of optometrists, ophthalmologists in assessing the child (for example in decompensated esophoria, congenital nystagmus) and understand the role of neurophysiology and OCT (as in intracranial hypertension) in investigations
7. Be able to differentiate central visual problems from pre-chiasmatic problems

Key activities

Essential

- Case log reflecting neuro-opthalmological problems in a neurological out patient clinics as well as in patient care
- Attend as observer a clinic of a paediatric optometrist and / or neuro-ophthalmologist

Desirable

- Attending case discussion/video discussion on neuroophthalmological problems
- Attending a regional national or international meeting on neuroophthalmological problems – may also be adult meeting
- Attend EPNS webinars on “Supranuclear eye movement problems” and “Development of the visual system – strabismus and nerve palsies”

16. Program on sleep disorders in children for a Trainee in Paediatric Neurology

Recognize normal and abnormal sleep behavior during infancy and childhood

Be able to recognize and manage the most frequently occurring sleep disorders in children (physiological problems such as adolescent delayed sleep onset disorder, obstructive sleep apnoea abnormal or disruptive behaviours during sleep such as sleepwalking or other parasomnias symptoms that occur near sleep onset such as restless legs syndrome, early childhood insomnias, and daytime symptoms such as excessive sleepiness, cataplexy , narcolepsy or Kleine Levin syndrome

Key activities

Essential

- Case log reflecting sleep problems in a neurological outpatient clinics as well as in patient care
- Attend rounds on sleep problems

Desirable

- Attend EPNS webinars " Up date on paediatric sleep problems" (as narcolepsy)"

17. Headache Programme for a Trainee in Paediatric Neurology

At the end of their training a trainee in paediatric neurology should be able to:

1. Undertake a detailed history and assessment of a child with suspected headache disorder (HD), including life style risk factors.

2. Be familiar with primary and secondary headache disorders, and be familiar with the ICDH-beta classification and the NICE guideline for headaches in children >12 years of age.

3. Understand the appropriate investigation for headaches and when no further investigations are necessary.

4. Diagnose and manage raised intracranial pressure.

5. Assess, investigate and diagnose a child presenting with common headache types including migraine.

6. Diagnose and manage common migraine variants and periodic syndromes.

7. Diagnose and manage patients with idiopathic intracranial hypertension (IIH), including investigations such as MRI/V, ophthalmic assessment including orbital coherence tomography / ultrasound, treatment with spinal taps, carbonic anhydrase inhibitors, weight reduction and surgical intervention

8. Lead on inter-disciplinary working with allied specialities for this condition including ophthalmology, psychology

<u>Acute neurology</u>

- Be familiar with the recognition, investigation and management of headache as a presentation of acutely raised ICP
- Be aware of the presentation, investigation and management of sub-arachnoid haemorrhage

<u>Transition and adult neurology</u>

- Understand adult headache disorders that can present in the childhood/adolescent period
- Be aware of the presentation, investigation and management of sub-arachnoid haemorrhage

Neuropsychological and neuropsychiatric aspects

- Understand and manage the impact of headaches on the child and family
- Know how to recognise and initiate management for chronic medically un-explained headache
- Be familiar with the role of psychological approaches in the management of chronic headache

Neurodisability

- Recognise the different presentation of headache disorders in a child with moderate to severe neurodisability
- Be confident in the assessment and management of headache in a child with neurodisability

Key activities

Trainees should be encouraged to participate in the following activities to support their training in headache disorders:

Essential:

- Reflective Case Log of 20 patients with primary and secondary headache disorders
- Case log of at least five cases where a child or young adult has been referred for assessment as to whether they have idiopathic intercranial hypertension
- Attendance at a national or regional meeting on Children's Headache

Desirable:

- Audit, service evaluation or quality improvement project in relation to headache disorders (could be adult)
- Attendance at a national or regional headache specialist interest group meeting
- Completion of Unit 12 of the British Distance Learning Programme
- Attendance of the EPNS webinar "Primary headaches" and "Dangerous headaches"

18. Acquired Brain Injury and Neurorehabilitation Programme for Trainee in Paediatric Neurology

At the end of their training a trainee in paediatric neurology should:

1. Be familiar with assessment, investigation and management of a child presenting with mild or complicated mild traumatic brain injury
   Be familiar with the initial assessment, investigation and management of a child presenting with severe traumatic and non-traumatic encephalopathy in a major trauma center with a neurosurgical ITU.
2. Have participated in MD discussions regarding prognosis and withdrawal of treatment, including brain death assessment. See also chapter on Diagnosis of brain death

3. Be familiar with 'early' management considerations and treatment ie within first few weeks of extuation.

4. Understand the role of assessment tools and scales to evaluate impairments, disability and quality of life in the ongoing rehabilitation of a child following acquired brain injury (ABI).

5. Be confident in providing medical input and contributing to multi-disciplinary goal planning meetings to determine on going management of a child with ABI including their educational, social and psychological well-being.

6. Be aware of factors that affect prognosis and long-term outcome in ABI.

7. Be confident in leading complex discharge planning meetings for a child with ABI and the importance of agreeing holistic goals.

8. Be familiar with referral pathways and multidisciplinary services for the management of children with rehabilitation needs. Be able to engage other key professionals in co-ordinating rehabilitation and care packages, linking with tertiary and primary health care, education, social services and the voluntary sector.

9. Be familiar with the assessment and management of children with spinal injury including the features and management of autonomic dysreflexia.

10. Be familiar with the characteristics of inflicted traumatic brain injury and the relevant safeguarding procedures and referral pathways.

11. Understand the importance of providing accurate information to other agencies and the need to update this information regularly as the child's needs changes. Be able to draft reports for other agencies including social services and the police.

Acute neurology

- Be confident in the acute assessment and management of a child with acute traumatic encephalopathy, presenting via a major trauma route, through ER or acutely deteriorating on the ward
- Be confident in the management of raised intracranial pressure and the role of neurosurgical intervention and pressure monitoring
- Be confident in the assessment of a child with suspected spinal injury, in particular the priorities in investigation and management
- Be aware of importance of early MD input in a child presenting with AB or spinal injury and be confident in liaising with critical care teams, ER and neurosurgical colleagues

Transition and adult neurology

- Be aware of the potential risk factors and needs of the patient and family presenting with an ABI in adolescence
- Be familiar with transitioning care of young people with ABI to adult services and the existing referral pathways and services

Neuropsychological and neuropsychiatric aspects

- Be aware of the pre-existing cognitive, behavioural and social risk factors for ABI and its outcome
- Recognise the common neuropsychological co-morbidities that can result from ABI or cord injury
- Understand the role of psychology/psychiatry/CAMHS services in the management of children with ABI or cord injury and their families and the indications for referral
- Understand the specific role of neuropsychiatric assessment in children with ABI

Neurodisability

- Be confident in the ability to ascertain pre-morbid developmental, behavioural, emotional, cognitive and physical difficulties
- Appreciate the significant increased risk of head injuries in children with other neurodevelopmental conditions
- Understand the importance of including a consideration of previous acquired brain injury in the work up of children with neurodevelopmental disorders.
- Understand the increased risk of delayed complications for children post ABI, eg seizures and headaches

Key activities

Trainees would be encouraged to participate during their training in acquired brain injury (ABI) and neurorehabiliation in the following acitvities:

Essential

- A case log of 10 severe traumatic and non-traumatic encephalopathy cases which should include reflective case examples of the following:
    - Observations of brain death assessment (1-2 cases)
    - Treatment withdrawal discussions with families (3 patients)
    - Case of possible abusive head trauma (1-2 cases)
- Case log detailing early acute progress (ie from extubation onwards for 1 -2 weeks) of 4 children with severe ABIs and 1 more detailed CBD
- Case log following the management of further 10 patients with ABI through their rehabilitation journey
- Have experience of working in a supra-regional rehabilitation facility and MTC or participate in MD follow up clinics
- Attend 3 x MDT relating to a child with ABI to observe interdisciplinary team working and goal setting
- Participate in, and lead at least 1 of 3 complex discharge planning meetings for a child undergoing neurorehabiliatation
- Observe and reflect on at least 2 neuropsychological assessments and preparation of educational advice
- Keep a case log of 10 children with spinal cord “injury” – including myelitis, tumour, trauma

Desirable

- Spend time in a specialist spinal problems out patient clinic or in patient unit (such as MMC, dysgraphia, injuries) - ideally paediatric but adult if necessary
- Attend Neurorehabilitation specialist interest meeting
- Undertake an audit or service evaluation that relates to children with ABI
- Case report or presentation of child with ABI
- Completion of British Distance Learning Unit 0 Introduction to Paediatric Neurology
- Aattend EPNS training course on Acute Neurology
- Attend EPNS webinars on “Acute head trauma” and “Reorganisation and Nuerorehabilitation- what do we know”

19. Program on Diagnosis of brain death and end of life care in Paediatric Neurology

At the end of their training a trainee in paediatric neurology should be:

1. Capable to diagnose brain death according to national guidelines. Being aware of complexity of the assessment and impact on families and supporting team.

2. Know about information on neurophysiological and imaging information on brain death

3. Be able to participate in assessing the life prognosis, in decisions on appropriate care and for symptom control in children who do not meet the criteria for brain death

4. Being able to discuss with parents of children with a severely disabling neurological condition and an expected limited life span which life sustaining therapies will or will not be started in case of an acute deterioration

Key activities

Essential

- Being closely involved with at least one assessment of brain death,
- Discussion of prognosis and treatment options in > 2 children with life limiting illness
- Attendance at a morbidity and mortality meeting
- Log of at least two cases with complex multidisciplinary input for end of life care

Desirable

- Watch video on assessment of brain death
- Attendance at an case discussion of a child with life limiting illness and/or brain death with an ethics team
- Case presentation on a child with difficult decision on continuing care
- Attendance of a debriefing meeting after a child`s death
- Attend EPNS webinars “Diagnosis of brain death” and “Prognostic markers for outcome in acute encephalopathies”

20. Medically Unexplained Neurological Disorders Programme for a Trainee in Paediatric Neurology

At the end of their training a trainee in paediatric neurology should:

1. Be aware of the symptoms and signs that might indicate a medically unexplained neurology disorders (MUND) across different age groups.

2. Be aware of the atypical presentations of other neurological illness that can erroneously be thought to have a 'non-organic' basis. In particular, movement disorders, headache, sensory abnormalities, inflammatory disorders and epilepsy.

3. Be able to take a detailed history and assessment of a child and family where MUND is suspected.

4. Be able to communicate the diagnosis to the child and family and other relevant professionals.

5. Understand the role of investigations and their potential negative impact in MUNDs.

6. Be aware of the fact that a child may have coincidental MUND and a neurological disorder ie non epileptic attacks in a child with a confirmed seizure disorder.

7. Be able to develop a plan for the ongoing management in a child with MUNDs.

8. Recognize factors that may affect prognosis in MUNDs.

9. Be familiar with referral pathways and multidisciplinary services for the management of children with MUNDs.

10. Be aware of parental/family actions that could result in a child presenting with MUNDs ie FII and how this should be approached and investigated.

Acute neurology

- Be aware of the potential acute presentation of MUNDs with status epilepticus/ paralysis/dystonia etc and the features that can help distinguish the two
- Be aware of importance of early MDT input and psychiatric advice in a child presenting acutely with MUNDs to avoid unnecessary investigations and treatment, especially when FII is suspected

Transition and adult neurology

- Be aware of the potential risk factors and needs of the patient and family presenting with a MUND in adolescence
- Be familiar with the support networks and strategies that exist to support young adults with MUND

Neuropsychological and neuropsychiatric aspects

- Be aware of the associated cognitive and behavioural risk factors for MUND
- Recognise the common neuropsychological co-morbidities that can result from MUND
- Understand the role of both neurology and psychology/psychiatry/CAMHS services in the management of children and their families when a diagnosis of MUND is made

Neurodisability

- Be aware of the symptoms and signs of MUND in a child with pre-existing neurodisability and the strategies required to help distinguish functional from organic symptoms
- Recognise the limitations/challenges of certain investigations ie prolonged EEG to investigate MUND in children with neurodisability
- Be able to formulate management strategies with families, working closely with the multidisciplinary team and other agencies

Key activities

Trainees should be encouraged to participate in the following activities to support their training in medically unexplained neurological disorders (MUND):

Essential

- Brief case log of 10 patients presenting acutely and in outpatients with a spectrum of different MUNDs
- Attendance at 10 psychology/psychiatry/CAMHS clinics during training
- Completion of 2 case based discussions during training that cover the assessment, acute and ongoing management of a child presenting with MUND, ideally in both out and inpatient settings
- Attendance at 1 complex MDT planning meeting for a child with diagnosis of MUND

Desirable

- Evidence of audit or service evaluation that relates to children with MUND
- Case report or presentation of child with MUND
- Attend EPNS teaching course “Acute Neurology”
- Attend EPNS webinar “Movement problems in MUND”

21. Program on Aspects of Adult Neurology in Paediatric Neurology

At the end of their training a trainee in paediatric neurology should able to do the below. It is likely that the competencies will be gained efficiently through exposure to adult neurology practice:

1. Being able to perform a neurological examination for localization of the relevant lesion/disease process within CNS as well as nerve/muscle problems
2. Being capable of a clinical neuroophthalmological examination for the purpose of localization / diagnosis (visual fields, oculomotor movements, fundoscopy, frenzel glasses, RAPD etc)
3. Being aware of the most important neurological problems in adult life, which do occur in children but exposure within training may be limited (e.g. cerebrovascular problems, multiple sclerosis, Parkinson/movement disorders, dementia, MUND etc) and being capable of performing a diagnostic investigation plan and knowing the difference of these problems and their diagnostic investigation to the same problems presenting already during childhood
4. Having an understanding for the most important neurophysiological and neuroimaging investigation in adulthood and knowing the differences to the same techniques in childhood.

Key activities

Essential

- Participating in outpatient clinics or inpatient care of adult neurology services e.g. cerebrovascular problems, multiple sclerosis, Parkinson/movement disorders, dementia, MUND
- Participating at different rounds with case discussion of adult neurology patients

Desirable

- Spending 3-6 month in an adult neurology service
- Participating at rounds and educational events for adult neurology
- Attend EPNS Webinars on “Clinical localization - a first approach” and “Adult disorders presenting in childhood and childhood disorders presenting in adulthood

Guidelines EPNS, adapted from the British Grid training program