

SCHEDULE 2 – THE SERVICES

A. Service Specifications

Service Specification No.	E09/S(HSS) tba
Service	Rare Hereditary Neuropathies for Children and Young People
Commissioner Lead	
Provider Lead	
Period	
Date of Review	

1. Population Needs

1.1 National/local context and evidence base

The service will be a highly specialised hub and spoke network of up to four expert centres which will provide comprehensive assessment, electrophysiological and genetic diagnosis and management for children and young people who have a suspected Rare Hereditary Neuropathy (RHN).

Children and young people will be referred in to the service following local diagnostics including testing for the four common genes is provided from any UKGTN approved molecular genetics laboratory. The service will add value to the patient's pathway by using the test results, interpreting these and creating a bespoke treatment package

The service will include: comprehensive multi-disciplinary clinical assessment; Deoxyribonucleic Acid (DNA) diagnosis; laboratory diagnosis of biopsies; any further genetic testing that is required; genetic counselling including antenatal testing and pre-implantation diagnosis; and access to generic and gene-specific therapies. It will also support improved training of multi-disciplinary clinicians.

The accurate diagnosis of these patients will avoid unnecessary or invasive tests and interventions, contributing to efficient use of resources.

Given that patients with these conditions frequently have co-morbidities, the provision of holistic care requires the involvement of multiple health professionals both in the community / local hospital services and in the tertiary setting. Currently most centres will have formally

established links between the tertiary centre and the regional community/ district general hospital setting which are central to the ability to provide a comprehensive package of care to children/young people and their families.

National context

The hereditary neuropathies are a clinically and genetically diverse group of neurological disorders that can affect children and young adults, causing degeneration of nerves in the peripheral nervous system (PNS). They are due to an inherited change in the genetic code in many different genes for synthesis of proteins involved in axonal or myelin function or survival. Some of these disorders are common, with over 90% of patients having one of four main types of hereditary neuropathy that are not challenging to diagnose. However the rarest types which affect the remaining 10% of patients are very difficult to diagnose and specialist testing is required, together with the expertise to interpret results and propose management plans to ensure appropriate management.

Most hereditary neuropathies present in childhood or adolescence and cause muscle wasting, weakness, hand and feet deformity, pain and significant disability. Some of them affect only the peripheral nerves and others affect the brain as well as other organs. Mutations in more than 80 different causative genes have been found so far, and the number of causative genes steadily increasing. Gene specific therapies are being trialled.

The RHN service is for the 10% of patients affected by the rare hereditary neuropathies and will provide a comprehensive multi-disciplinary clinical assessment, DNA diagnosis, accurate genetic counselling including antenatal testing and pre-implantation diagnosis, access to generic and gene-specific therapies.

The accurate diagnosis of these patients will avoid unnecessary or invasive tests and expensive interventions, contributing to efficient use of resources. This will include optimal medical, physiotherapy, orthotic and orthopaedic management, realistic goal setting and understanding of disease specific complications.

2. Outcomes

2.1 NHS Outcomes Framework Domains & Indicators

Domain 1	Preventing people from dying prematurely	✓
Domain 2	Enhancing quality of life for people with long-term conditions	✓
Domain 3	Helping people to recover from episodes of ill-health or following injury	✓
Domain 4	Ensuring people have a positive experience of care	✓
Domain 5	Treating and caring for people in a safe environment and protecting them from	✓

Outcomes

Domain 1: Preventing people from dying prematurely

Improvement area: Rare treatable conditions misdiagnosed as 'hereditary neuropathy' due to metabolic or acquired inflammatory disorders may receive the right treatment and hence morbidity and mortality averted.

Occasional metabolic inherited neuropathies that present as a Charcot-Marie-Tooth (CMT)-like syndrome are treatable, such as Brown Vialetto Van Laere syndrome (BVVL) if diagnosed.

Prompt diagnosis and treatment can prevent early death.

Domain 2: Enhancing Quality of Life for people with long term health conditions.

Giving patients precise diagnosis helps them come to terms with their chronic medical condition.

Signposting them to right treatment and other useful interventions will improve physical and psychosocial outcomes.

Genetic counselling and where appropriate antenatal diagnosis will help patients make appropriate reproductive choices.

Domain 3: Helping people to recover from episodes of ill-health or following injury.

In instances of hereditary neuropathy which were previously misdiagnosed as acquired inflammatory neuropathies and treated with high doses of expensive and/or toxic drugs, appropriate genetic diagnosis will lead to discontinuation of unnecessary therapies with reduced costs and positive consequences for the patient.

Domain: 4 Ensuring that people have a positive experience of care

As above

Overarching indicator:

- Patient right to a correct diagnosis
- Patient experience of hospital care

Improvement area:

- Patient experience of outpatient services

This service specification will ensure that patients receive care through a national clinical network.

Service providers will provide outcomes data on:

- Patient experience of outpatient services through a patient questionnaire developed and validated with appropriate patient representative groups
- Number of people who are referred to specialist care

- Number of patients initiated on approved treatments

Domain 5: Treating and caring for people in a safe environment and protecting them from avoidable harm

Overarching indicator:

- Patient safety incidents reported
- Safety incidents involving severe harm or death
- Hospital deaths attributable to problems in care

3. Scope

3.1 Aims and objectives of service

Aim: The provision of a dedicated multidisciplinary service for children and young adults with rare hereditary neuropathies.

Objectives: Improved access to clinical expertise and emerging treatments including reproductive planning for patients with rare hereditary peripheral neuropathies.

3.2 Service description/care pathway

The proposed dedicated multidisciplinary services for rare hereditary neuropathies will be provided via a network at up to four expert centres in England. Access will be available to chemical pathology, neuropathology and molecular genetics laboratories.

The minimal prevalence of hereditary peripheral neuropathies is 11 per 100,000 in Northern England across all ages. Figures as high 40-80 per 100,000 have been quoted in other papers. The minimal estimated total number of patients with hereditary peripheral neuropathies in England (population of 53 million) is 6300. This includes the four common gene mutations plus the rare ones.

The National centre will be for the rare hereditary neuropathies which may account for 10% of the total, which gives an overall population of 630 patients, of which the paediatric population is expected to be some 50% or 300 patients in total.

It is estimated that up to 100 patients will be assessed per year to include all new patients initially and with a new to follow up ratio of 50:50 annually thereafter. The remainder of the caseload of patients will be managed locally following initial assessment, diagnosis and creation of care plans.

Patients seen at their local centre who are suspected to be affected by a hereditary neuropathy, will have basic tests including molecular genetics for mutations in the four common genes that currently account for up to 90% of the UK patient population with a genetic defect, and, if appropriate, electrophysiology. If these tests are negative or

inconclusive they will be referred to one of the dedicated neuropathy centres for further evaluation and investigations, and for management advice.

Diagnostics

Comprehensive molecular genetic testing for individuals suspected to be affected by a rare hereditary neuropathy will be available via the dedicated centres to ensure appropriate gate keeping that will result in an overall reduction in cost associated with the investigation of rare hereditary neuropathies.

While next generation sequencing techniques are rapidly developing and it is envisaged will soon be available to also cover genes responsible for rare genetic neuropathies, the need of expert interpretative skills will, if anything, increase in the future as scenarios of patients with variants of unknown significance in multiple genes is a likely outcome.

Knowledge of the function of individual genes, and of the electrophysiological and clinical consequences of their dysfunction will be required to properly assign pathogenicity to these changes. Also many mutations in the rare genes are novel and a service to clinically evaluate (including neurophysiology) affected and unaffected relatives to establish segregation is crucial as segregation of a novel mutation with a disease is still the most powerful tool in establishing pathogenicity of a mutation.

Gene-specific therapies if available in the future will be delivered through the centres liaising with clinical pharmacists.

Generic rehabilitative therapies will be delivered through liaison with the patient's community teams.

Patients will be monitored once or twice yearly depending on need in the joint dedicated multidisciplinary clinics which will be held at the respective centres. These will be one stop clinics in a multidisciplinary setting and will include assessments on the day which will be undertaken by the neurologist, orthopaedic surgeon, physiotherapist, occupational therapist, orthotist and neuromuscular care advisors. There will be close liaison with the local referring teams and the community services, facilitated by the family care liaison advisers. Expert advice on treatment and management of patients with an established rare neuropathy will be provided as well if requested.

Investigations

Required investigations or monitoring such as radiology will be available on the day by CPA accredited services. Biochemistry, haematology and molecular genetics samples will be taken on the day of the clinic, sent to the appropriate laboratory and the family will be invited back to clinic for an assessment of the results and for discussions on a proposed package of care based on this.

Additional allied specialists such as speech and language therapists or psychologists will be available as required. Neuroradiology (brain, spinal cord, nerve and muscle MRI) will be also available at the individual centres, together with expertise in the interpretation of findings in the context of a rare neuropathy. Dedicated centres will have access to gait analysis service if needed.

Staffing

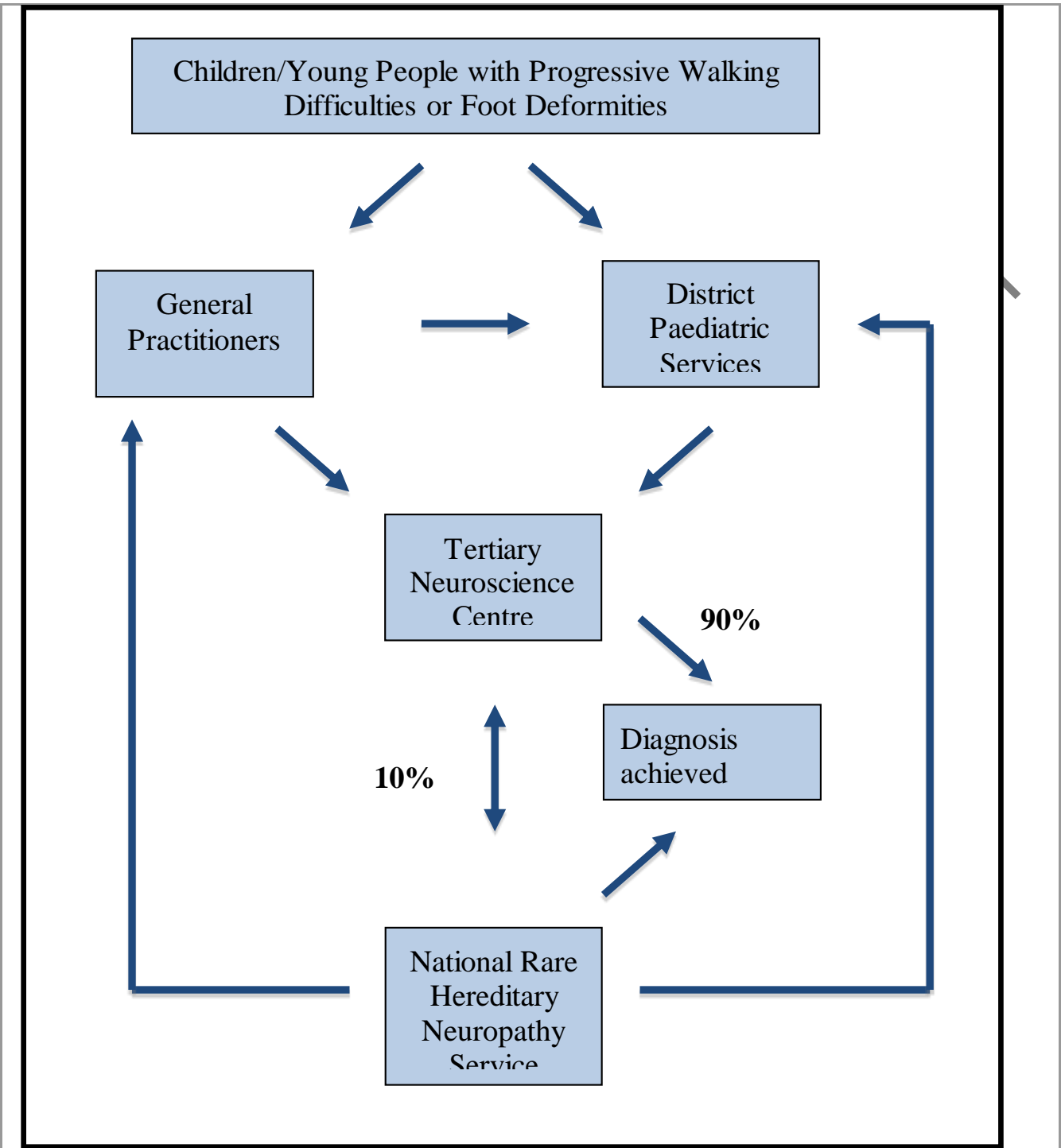
The multi-disciplinary teams will include:

- paediatric and adult neurologists,
- specialised electrophysiology,
- orthopaedic surgeons,
- physiotherapists,
- occupational therapists,
- orthotists,
- psychologists and neuromuscular care advisors.

The clinical team will have access to expert opinion on neuroradiology, metabolic medicine, neurosurgery, neuropathology, neurogenetics as required.

Model of care

There will be a hub and spoke model of care with shared care arrangements between local services and specialist centres. This will include alternate clinic appointments to reduce travelling and to ensure that as much of the care as possible will be delivered locally. Clinical care pathways for individuals affected by rare hereditary neuropathies will be developed.



Education and Teaching

Regular regional teaching will be provided to primary, secondary and tertiary teams involved in the care of this patient group who have complex and often challenging conditions.

Registries

Patient registries will be established to provide accurate data on patient numbers and diagnosis which will facilitate research and provide information for future clinical trials.

Shared learning

There will be regular national meetings of participating centres for clinical case discussion and reviews. These meetings will be performed in collaboration with the British Peripheral Nerve society, British Paediatric Neurology Association and Association of British Neurologists.

Transition

The dedicated centres will formalise transitional care arrangements in collaboration with the respective adult services. Joint transitional clinics will be established.

Documentation and Information Technology

Following each multidisciplinary clinic, the staff will produce a single clinic letter which will integrate the reports from each clinician. This will be sent out to the patient and their family or carer; the patient's GP/and/or specialist.

RHN service clinic notes and correspondence will be stored at each centre (computerised) and included in the hospital-wide computerised patient records of the patient. The provider local administrator will have responsibility for ensuring safe storage and adherence to the Data Protection Act (1998) for computerised data. Offsite backup storage will also be arranged.

Overview

Patient Referral

Patients seen at their local centre who are suspected to be affected by a hereditary neuropathy will have basic tests including molecular genetics for the mutations in the four common genes that currently account for up to 90% of the UK patient population with a genetic defect, and, if appropriate, electrophysiology. If these tests are inconclusive, they will be referred to one of the dedicated neuropathy centre for further evaluation and investigations, and for management advice.

Discharge criteria

Following initial attendance and diagnostic assessment, if the patient does not meet entry criteria, they will be discharged back to local tertiary care providers.

Outpatient service requirements: Existing joint multi-disciplinary outpatient clinic provision at designated centres.

Inpatient service requirements: This will be rarely needed.

Risk management

Services delivered by the RHN service providers must be of a nature and quality to meet the standards set out in this specification. It is the provider Trust's responsibility to notify the

commissioner on an exceptional basis should there be any breaches of the standards. Where there are breaches, any consequences will be deemed as being the Trust's responsibility.

Equity of access to services

Access to the service will be paramount to its success. It is not expected that all patients will need to travel to specialised centres and a tiered service will be offered related to the needs of the individual.

3.3 Population covered:

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

http://www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH_127393)

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

3.4 Any acceptance and exclusion criteria and thresholds

The service will accept referrals of patients from birth to 21st birthday with a suspected or proven Rare Hereditary Neuropathy (RHN).

3.5 Interdependencies with other services/providers

The Clinical Network must be able to demonstrate that it has well established links or an agreed plan, to:

- Primary care.
- A full range of diagnostic imaging, clinical neurophysiology and pathology services.
- Formal links to other specialist centres to facilitate transfer of care as required
- Formal arrangements to support continuation of treatment of patients moving between or leaving secure environments
- Formal pathways to support transition of paediatric patients to adult services as required
- Mental health services for patients with significant mental health needs – ranging from third sector support services to clinical psychology, liaison psychiatry and liaison with community mental health services in patients' place of residence.
- The Network will ensure that it describes the links and interfaces of its services and care with other relevant pathways and organisations (e.g. Local Authorities) as required.

4. Applicable Service Standards

4.1 Applicable national standards e.g. NICE

Currently there are no national standards for the rare peripheral neuropathies. The hub centres will collaboratively develop service standards as a priority.

4.2 Applicable standards set out in Guidance and/or issued by a competent body

Currently there are no published standards for the rare peripheral neuropathies. The hub centres will collaboratively develop service standards as a priority.

4.3 Applicable local standards

The providers of the service must ensure they are fully integrated into their Trust's corporate and clinical governance arrangements and must comply fully with clinical negligence scheme for trusts (CNST) and Care Quality Commission (CQC) requirements in terms of quality and governance.

5.1 Applicable quality requirements (See Schedule 4 Parts A-D)

There is a requirement to hold national audit meetings at their centre and involving all designated centres on an annual basis as a minimum. Any service issues should be flagged to the specialist commissioning Hub contracting team and the Highly Specialised Services Team.

Each centre must assure that:-

- All practitioners participate in continuous professional development and networking
- Patient outcome data is recorded and audited across the service
- All centres must participate in the national audit commissioned by NHS England.

Audit meetings will address:

- Clinical performance and outcome
- Process-related indicators e.g. efficiency of the assessment process, prescribing policy, bed provision and occupancy, outpatient follow-up etc.
- Stakeholder satisfaction, including feedback

5.2 Applicable CQUIN goals (See Schedule 4 Part E)

To be agreed with the commissioner.

6. Location of Provider Premises
To be agreed. Centres and linked services will be defined through the compliance process
7. Individual Service User Placement
Not Applicable

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Glossary of Terms		
BVL	Brown Vialto Van Laere syndrome	A disorder which is characterised by nerve problems, particularly hearing loss caused by nerve damage in the inner ear (sensorineural hearing loss) see http://ghr.nlm.nih.gov/condition/brown-vialto-van-laere-syndrome
CMT	Charcot-Marie-Tooth disease	A group of inherited disorders of the peripheral nervous system. Clinical diagnosis can be made fairly accurately, and the clinical classification is very helpful in the identification of new disease genes.
CNST	Clinical Negligence Scheme for Trusts	http://www.nhs.uk/claims/Documents/CNST%20Rules.pdf
CQC	Care Quality Commission	The independent regulator of health and social care in England http://www.cqc.org.uk/
UKGTN	UK Genetic Testing Network	The UK Genetic Testing Network is an advisory organisation that provides commissioning support to the NHS and DH for NHS patients in the UK http://ukgtn.nhs.uk/
PNS	Peripheral Nervous System	Peripheral nervous system: The portion of the nervous system that is outside the brain and spinal cord.
RHN	Rare Hereditary Neuropathy(ies)	Hereditary neuropathies are a group of inherited disorders affecting the peripheral nervous system

References

J Neurol Neurosurg Psychiatry 2012 83: 572-573

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Appendix One

PROVISION OF SERVICES TO CHILDREN

Aims and objectives of service

This specification annex applies to all children's services and outlines generic standards and outcomes that would be fundamental to all services.

The generic aspects of care:

The Care of Children in Hospital (Health Service Circular 1998/238) requires that:

- Children are admitted to hospital only if the care they require cannot be as well provided at home, in a day clinic or on a day basis in hospital.
- Children requiring admission to hospital are provided with a high standard of medical, nursing and therapeutic care to facilitate speedy recovery and minimize complications and mortality.
- Families with children have easy access to hospital facilities for children without needing to travel significantly further than to other similar amenities.
- Children are discharged from hospital as soon as socially and clinically appropriate and full support provided for subsequent home or day care.
- Good child health care is shared with parents/carers and they are closely involved in the care of their children at all times unless, exceptionally, this is not in the best interest of the child. Accommodation is provided for them to remain with their children overnight if they so wish.

Service description/care pathway

All paediatric specialised services have a component of primary, secondary, tertiary and even quaternary elements.

The efficient and effective delivery of services requires children to receive their care as close to home as possible dependent on the phase of their disease.

Services should therefore be organised and delivered through "integrated pathways of care" (National Service Framework for children, young people and maternity services (Department of Health & Department for Education and Skills, London 2004))

Interdependencies with other services

All services will comply with *Commissioning Safe and Sustainable Specialised Paediatric Services: A Framework of Critical Inter-Dependencies* – Department of Health (DH)

Imaging

All services will be supported by a 3 tier imaging network ('Delivering quality imaging services for children' DH 13732 March 2010). Within the network:

- It will be clearly defined which imaging test or interventional procedure can be performed and reported at each site

- Robust procedures will be in place for image transfer for review by a specialist radiologist, these will be supported by appropriate contractual and information governance arrangements
- Robust arrangements will be in place for patient transfer if more complex imaging or intervention is required
- Common standards, protocols and governance procedures will exist throughout the network.
- All radiologists, and radiographers will have appropriate training, supervision and access to continuing professional development
- All equipment will be optimised for paediatric use and use specific paediatric software.

Specialist Paediatric Anaesthesia

Wherever and whenever children undergo anaesthesia and surgery, their particular needs must be recognised and they should be managed in separate facilities, and looked after by staff with appropriate experience and training.¹ All UK anaesthetists undergo training which provides them with the competencies to care for older babies and children with relatively straightforward surgical conditions and without major co-morbidity. However those working in specialist centres must have undergone additional (specialist) training² and should maintain the competencies so acquired^{3*}. These competencies include the care of very young/premature babies, the care of babies and children undergoing complex surgery and/or those with major/complex co-morbidity (including those already requiring intensive care support).

As well as providing essential co-dependent service for surgery specialist anaesthesia and sedation services may be required to facilitate radiological procedures and interventions (for example MRI scans and percutaneous nephrostomy) and medical interventions (for example joint injection and intrathecal chemotherapy), and for assistance with vascular access in babies and children with complex needs such as intravenous feeding.

Specialist acute pain services for babies and children are organised within existing departments of paediatric anaesthesia and include the provision of agreed (hospital wide) guidance for acute pain, the safe administration of complex analgesia regimes including epidural analgesia, and the daily input of specialist anaesthetists and acute pain nurses with expertise in paediatrics.

*The Safe and Sustainable reviews of paediatric cardiac and neuro- sciences in England have noted the need for additional training and maintenance of competencies by specialist anaesthetists in both fields of practice.

References

1. Guidelines for the Provision of Anaesthetic Services (GPAS) Paediatric anaesthetic services. RCoA 2010 www.rcoa.ac.uk
2. Certificates of Completion of Training (CCT) in Anaesthesia 2010
3. Continuing Professional Development (CPD) matrix level 3

Specialised Child and Adolescent Mental Health Services (CAMHS)

The age profile of children and young people admitted to specialised CAMHS day/in-patient settings is different to the age profile for paediatric units in that it is predominantly adolescents who are admitted to specialised CAMHS in-patient settings, including over-16s. The average length of stay is longer for admissions to mental health units. Children and young people in specialised CAMHS day/in-patient settings generally participate in a structured programme of education and therapeutic activities during their admission.

Taking account of the differences in patient profiles the principles and standards set out in this specification apply with modifications to the recommendations regarding the following:

- Facilities and environment – essential Quality Network for In-patient CAMHS (QNIC) standards should apply (<http://www.rcpsych.ac.uk/quality/quality,accreditationaudit/qnic1.aspx>)
- Staffing profiles and training - essential QNIC standards should apply.
- The child/ young person's family are allowed to visit at any time of day taking account of the child / young persons need to participate in therapeutic activities and education as well as any safeguarding concerns.
- Children and young people are offered appropriate education from the point of admission.
- Parents/carers are involved in the child/young person's care except where this is not in the best interests of the child / young person and in the case of young people who have the capacity to make their own decisions is subject to their consent.
- Parents/carers who wish to stay overnight are provided with accessible accommodation unless there are safeguarding concerns or this is not in the best interests of the child/ young person.

Applicable national standards e.g. NICE, Royal College

Children and young people must receive care, treatment and support by staff registered by the Nursing and Midwifery Council on the parts of their register that permit a nurse to work with children (Outcome 14h Essential Standards of Quality and Safety, Care Quality Commission, London 2010)

- There must be at least two Registered Children's Nurses (RCNs) on duty 24 hours a day in all hospital children's departments and wards.
- There must be an Registered Children's Nurse available 24 hours a day to advise on the nursing of children in other departments (this post is included in the staff establishment of 2RCNs in total).

Accommodation, facilities and staffing must be appropriate to the needs of children and separate from those provided for adults. All facilities for children and young people must comply with the Hospital Build Notes HBN 23 Hospital Accommodation for Children and Young People NHS Estates, The Stationary Office 2004.

All staff who work with children and young people must be appropriately trained to provide care, treatment and support for children, including Children's Workforce

Development Council Induction standards (Outcome 14b Essential Standards of Quality and Safety, Care Quality Commission, London 2010).

Each hospital who admits inpatients must have appropriate medical cover at all times taking account of guidance from relevant expert or professional bodies (National Minimum Standards for Providers of Independent Healthcare, Department of Health, London 2002). "Facing the Future" Standards, Royal College of Paediatrics and Child Health.

Staff must carry out sufficient levels of activity to maintain their competence in caring for children and young people, including in relation to specific anaesthetic and surgical procedures for children, taking account of guidance from relevant expert or professional bodies (Outcome 14g Essential Standards of Quality and Safety, Care Quality Commission, London 2010).

Providers must have systems in place to gain and review consent from people who use services, and act on them (Outcome 2a Essential Standards of Quality and Safety, Care Quality Commission, London 2010). These must include specific arrangements for seeking valid consent from children while respecting their human rights and confidentiality and ensure that where the person using the service lacks capacity, best interest meetings are held with people who know and understand the person using the service. Staff should be able to show that they know how to take appropriate consent from children, young people and those with learning disabilities (Outcome 2b) (Seeking Consent: working with children Department of Health, London 2001).

Children and young people must only receive a service from a provider who takes steps to prevent abuse and does not tolerate any abusive practice should it occur (Outcome 7 Essential Standards of Quality and Safety, Care Quality Commission, London 2010 defines the standards and evidence required from providers in this regard). Providers minimise the risk and likelihood of abuse occurring by:

- Ensuring that staff and people who use services understand the aspects of the safeguarding processes that are relevant to them
- Ensuring that staff understand the signs of abuse and raise this with the right person when those signs are noticed.
- Ensuring that people who use services are aware of how to raise concerns of abuse.
- Having effective means to monitor and review incidents, concerns and complaints that have the potential to become an abuse or safeguarding concern.
- Having effective means of receiving and acting upon feedback from people who use services and any other person.
- Taking action immediately to ensure that any abuse identified is stopped
- and suspected abuse is addressed by:
 - having clear procedures followed in practice, monitored and reviewed that take account of relevant legislation and guidance for the management of alleged abuse
 - separating the alleged abuser from the person who uses services and others who may be at risk or managing the risk by removing the opportunity for abuse to occur, where this is within the control of the provider

- reporting the alleged abuse to the appropriate authority
- reviewing the person's plan of care to ensure that they are properly supported following the alleged abuse incident.
- Using information from safeguarding concerns to identify non-compliance, or any risk of non-compliance, with the regulations and to decide what will be done to return to compliance.
- Working collaboratively with other services, teams, individuals and agencies in relation to all safeguarding matters and has safeguarding policies that link with local authority policies.
- Participates in local safeguarding children boards where required and understand their responsibilities and the responsibilities of others in line with the Children Act 2004.
- Having clear procedures followed in practice, monitored and reviewed in place about the use of restraint and safeguarding.
- Taking into account relevant guidance set out in the Care Quality Commission's Schedule of Applicable Publications
- Ensuring that those working with children must wait for a full CRB disclosure before starting work.
- Training and supervising staff in safeguarding to ensure they can demonstrate the competences listed in Outcome 7E of the Essential Standards of Quality and Safety, Care Quality Commission, London 2010

All children and young people who use services must be:

- Fully informed of their care, treatment and support.
- Able to take part in decision making to the fullest extent that is possible.
- Asked if they agree for their parents or guardians to be involved in decisions they need to make.

(Outcome 4I *Essential Standards of Quality and Safety*, Care Quality Commission, London 2010)

Key Service Outcomes

Evidence is increasing that implementation of the national Quality Criteria for Young People Friendly Services (Department of Health, London 2011) have the potential to greatly improve patient experience, leading to better health outcomes for young people and increasing socially responsible life-long use of the NHS.

Implementation is also expected to contribute to improvements in health inequalities and public health outcomes e.g. reduced teenage pregnancy and STIs, and increased smoking cessation. All providers delivering services to young people should be implementing the good practice guidance which delivers compliance with the quality criteria.

Poorly planned transition from young people's to adult-oriented health services can be associated with increased risk of non-adherence to treatment and loss to follow-up, which can have serious consequences. There are measurable adverse consequences in terms of morbidity and mortality as well as in social and educational outcomes. When children and young people who use paediatric services are moving to access adult services (for

example, during transition for those with long term conditions), these should be organised so that:

- All those involved in the care, treatment and support cooperate with the planning and provision to ensure that the services provided continue to be appropriate to the age and needs of the person who uses services.

The National Minimum Standards for Providers of Independent Healthcare, (Department of Health, London 2002) require the following standards:

- **A16.1** Children are seen in a separate out-patient area, or where the hospital does not have a separate outpatient area for children, they are seen promptly.
- **A16.3** Toys and/or books suitable to the child's age are provided.
- **A16.8** There are segregated areas for the reception of children and adolescents into theatre and for recovery, to screen the children and adolescents from adult patients; the segregated areas contain all necessary equipment for the care of children.
- **A16.9** A parent is to be actively encouraged to stay at all times, with accommodation made available for the adult in the child's room or close by.
- **A16.10** The child's family is allowed to visit him/her at any time of the day, except where safeguarding procedures do not allow this
- **A16.13** When a child is in hospital for more than five days, play is managed and supervised by a qualified Hospital Play Specialist.
- **A16.14** Children are required to receive education when in hospital for more than five days; the Local Education Authority has an obligation to meet this need and are contacted if necessary.
- **A18.10** There are written procedures for the assessment of pain in children and the provision of appropriate control.

All hospital settings should meet the Standards for the Care of Critically Ill Children (Paediatric Intensive Care Society, London 2010).

There should be age specific arrangements for meeting Regulation 14 of the Health and Social Care Act 2008 (Regulated Activities) Regulations 2010. These require:

- A choice of suitable and nutritious food and hydration, in sufficient quantities to meet service users' needs
- Food and hydration that meet any reasonable requirements arising from a service user's religious or cultural background
- Support, where necessary, for the purposes of enabling service users to eat and drink sufficient amounts for their needs
- For the purposes of this regulation, "food and hydration" includes, where applicable, parenteral nutrition and the administration of dietary supplements where prescribed
- Providers must have access to facilities for infant feeding, including facilities to support breastfeeding (Outcome 5E, of the Essential Standards of Quality and Safety, Care Quality Commission, London 2010)

All paediatric patients should have access to appropriately trained paediatric trained dieticians, physiotherapists, occupational therapists, speech and language therapy, psychology, social work and CAMHS services within nationally defined access standards.

All children and young people should have access to a professional who can undertake an assessment using the Common Assessment Framework and access support from social

care, housing, education and other agencies as appropriate.

All registered providers must ensure safe use and management of medicines, by means of the making of appropriate arrangements for the obtaining, recording, handling, using, safe keeping, dispensing, safe administration and disposal of medicines (Outcome 9 Essential Standards of Quality and Safety, Care Quality Commission, London 2010). For children, these should include specific arrangements that:

- They are supported to have a health action plan
 - Facilities meet the appropriate requirements of the Disability Discrimination Act 1995
- They meet the standards set out in Transition: getting it right for young people. Improving the transition of young people with long-term conditions from children's to adult health services. Department of Health, 2006, London.

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Appendix Two

Quality standards specific to the service:

Quality Requirement	Threshold	Method of Measurement	Consequence of breach
Domain 1: Preventing people dying prematurely			
Rare treatable conditions misdiagnosed as 'hereditary neuropathy' due to metabolic or acquired inflammatory disorders may receive the right treatment and hence morbidity and mortality averted.	>80%	National database	To be addressed in annual service audit meeting
Domain 2: Enhancing the quality of life of people with long-term conditions			
<p>Giving patients precise diagnosis helps them come to terms with their chronic medical condition.</p> <p>Genetic counselling and where appropriate antenatal diagnosis will help patients make appropriate reproductive choices.</p>	tbc tbc	National database	To be addressed in annual service audit meeting
Domain 3: Helping people to recover from episodes of ill-health or following injury			
In instances of previously misdiagnosed acquired inflammatory disorders, appropriate immunomodulatory therapies will lead to recovery from episodes of ill health.	Tbc	Local database	To be addressed in annual service audit meeting

Quality Requirement	Threshold	Method of Measurement	Consequence of breach
Domain 4: Ensuring that people have a positive experience of care			
Patient experience of outpatient services through a patient questionnaire developed and validated with appropriate patient representative groups	Tbc	Local database	To be addressed in annual service audit meeting
Number of people who are referred to specialist care	Tbc		
Number of patients who initiate approved treatments	Tbc		
Domain 5: Treating and caring for people in a safe environment and protecting them from avoidable harm			
Patients initiating therapy will have been discussed at a multi-disciplinary meeting with documentation of the recommendations provided to the patient and the general practitioner	> 85% of patients	Local database	To be addressed in annual service audit meeting