

A09/S/c

**2013/14 NHS STANDARD CONTRACT  
FOR CARDIOLOGY: INHERITED CARDIAC CONDITIONS (ALL AGES)**

**PARTICULARS, SCHEDULE 2- THE SERVICES, A- SERVICE SPECIFICATIONS**

<b>Service Specification</b>	
<b>No</b>	A09/S/c
<b>Service</b>	Cardiology: Inherited Cardiac Conditions (All Ages)
<b>Commissioner Lead</b>	
<b>Provider Lead</b>	
<b>Period</b>	12 months
<b>Date of Review</b>	

## 1. Population Needs

### 1.1 National/local context and evidence base

Inherited Cardiac Conditions (ICCs) are a group of largely monogenic disorders affecting the heart, its conducting *system* and vasculature. The first indication is sometimes sudden cardiac death (SCD) often in adolescents or early adulthood. When an ICC is diagnosed there are implications for the relatives.

The last decade has seen dramatic advances in our understanding of the molecular pathology of ICCs. More than 50 ICCs have been recognised and genetics tests are increasingly available for the more common disorders such as hypertrophic cardiomyopathy (HCM) and for some rarer disorders, for example, Marfan syndrome and long QT syndrome (LQTS). The conditions are highly heterogeneous, both genetically and clinically.

Epidemiological evidence is incomplete, but suggests a combined total prevalence for ICCs of about 340,000 in the UK (this includes approximately 120,000 individuals affected by familial hypercholesterolaemia or FH). Risks associated with these conditions are highly variable, depending on the mutation and the spectrum of clinical risk factors. The average annual risk of SCD is about 0.1% for LQTS, and the annual mortality for HCM about 0.3-1%. Risks are substantially higher for patients with the most severe symptoms or in those who have experienced a resuscitated cardiac arrest. Mutation carriers with no identifiable clinical risk factors have lower absolute risks. For example Marfan syndrome can be fatal by age 30 or younger if untreated. An analysis of incidence and prevalence can be seen in Appendix 1.

Two prospective studies based on coroners' reports have estimated the incidence (in England) of sudden deaths that are unexplained and may be due to arrhythmia syndromes. These are sudden deaths (often known as SADS, or sudden arrhythmic death syndromes) where the heart appears normal at post mortem examination. Bowker (2003) estimated 0.5 sudden unexplained deaths/ 100,000 / year in adult Caucasians aged 16–64, with an estimated 143 SADS deaths per year in England. Behr (2007) suggested that there was significant under-recording and mis-recording of SADS deaths and estimated the rate as up to 1.34 deaths / 100,000 / year in Caucasians aged 4–64, and 544 annual deaths in England. Subsequent familial studies have suggested that up to half of these SADS deaths are explained by ICCs, especially LQT syndrome and Brugada syndrome and some subtle cardiomyopathies (Behr, 2003 and 2008). This proportion is likely to increase significantly as novel genetic disorders come to light.

Due to the complex and diverse ways in which ICC services are currently delivered, it is difficult to provide accurate data on current average caseloads and the annual throughput of services. This, coupled with uncertainty regarding the prevalence and incidence of ICCs, make it difficult to estimate the capacity required for the future. However, it is clear that provision is inadequate.

The scope of ICC services includes the following main categories of ICCs:

**Arrhythmia syndromes** caused by mutations in the proteins involved in generating the action potential; these are mainly the proteins making up sodium, potassium or calcium conducting channels in the membranes of the cardiac myocytes, but also some proteins that affect ion conduction indirectly. The arrhythmia syndromes include conditions such as LQT syndrome, short- QT syndrome (Morita 2008), Brugada syndrome and catecholaminergic polymorphic ventricular tachycardia (CPVT).

**Cardiomyopathies**, caused mainly by mutations in the proteins making up the contractile system of the myofibrils, such as actin, myosin and troponin. The cardiomyopathies include hypertrophic cardiomyopathy (HCM) characterised by asymmetrical thickening of the heart muscle, dilated cardiomyopathy (DCM), which weakens the heart muscle resulting in heart failure, and arrhythmogenic right ventricular cardiomyopathy (ARVC), which is a primarily arrhythmogenic heart muscle disorder. These conditions can lead to heart failure, stroke and arrhythmia.

**Inherited arteriopathies**, which cause catastrophic rupture of the blood vessels in addition to affecting other organs. These conditions include Marfan syndrome, caused by mutations in the protein fibrillin-1 (a component of connective tissue) (Dean 2007), Ehlers-Danlos syndrome<sup>i</sup> and Loeys-Dietz syndrome. Many other cases of aortic dissection and aneurysm are genetically based and familial. Some cases of aortic dissection relate to bicuspid aortic valve, which has a familial tendency.

**Muscular dystrophies**, a group of multi-system genetic disorders that cause

progressive muscle weakness and death of muscle cells. Some muscular

dystrophies affect the heart, leading to arrhythmias. Examples include Emery-Dreifuss muscular dystrophy and myotonic dystrophy.

**Families afflicted by Sudden Arrhythmic Death Syndrome (SADS)**, many of which will be found to have an inherited cardiac condition

Heterozygous familial hypercholesterolaemia (FH) is **not** included in the scope of this service specification as services for FH are commissioned by Clinical Commissioning Groups (CCGs). However, FH is considered an ICC. NHS England commissions services for patients with homozygous familial hypercholesterolaemia. It is caused by mutations in one of three genes involved in the uptake of cholesterol-rich low density lipoprotein into cells. This results in severe elevation of the blood levels of cholesterol and low density lipoprotein, leading to a high risk of premature coronary atherosclerosis (Austin 2004). There is therefore an obvious opportunity for CCGs to consider commissioning HFH via their regional ICC service. This model brings together expertise in ICCs along with the associated genetic testing, counselling and family evaluation, which is required in FH just as in other ICCs. It also brings the patient under the care of a cardiologist, given that coronary artery disease is the major fatal complication of this disease.

The PHG Foundation (Foundation for Genomics and Population Health) Report provides the following statement:

*‘Overall in the UK the capacity of services is inadequate to meet either current or future estimated needs. Service provision is highly unequal in quality as well as quantity across the country. Estimates based on extrapolation from current service provision suggest that services would need to see an additional 7,000 new patients per year to bring the most poorly provided services (mostly in the regions) up to the level of the best. Epidemiological data suggest that the true shortfall may be significantly higher and that it will be likely to increase as more genetic tests become available and cascade testing gathers pace.....service provision needs to be increased across the UK including London.’*

### Specific causes

Condition	International prevalence	UK prevalence	Estimated prevalent UK cases (based on entire population 60980000)	Estimated prevalent YH cases (based on entire population 5278800)
Hypertrophic cardiomyopathy	1 in 500 Europe		121,960	10558
Arrhythmogenic right ventricular cardiomyopathy	1 in 1,000 to 1 in 10,000 European		6,098–60,980	528-5279

Long QT syndrome	1 in 5,000 European	1 in 2,000 to 1 in 5,000	12,196– 30,490	1056-2639
Dilated cardiomyopathy	36.5* in 100,000 USA		5,565*	1927*
Marfan syndrome	1 in 5,000 Worldwide	1 in 5,000	12,196	1056
Brugada syndrome	146.2 in 100,000 Japan  1 in 5,000 European		12,196	1056
Myotonic dystrophy	1 in 8,000 worldwide		7,623	660
Catecholaminergic polymorphic ventricular tachycardia	1 in 10,000 Europe		6,098	528
Ehlers-Danlos syndrome type IV	1 in 250,000 USA		244	21
Noonan syndrome	Birth prevalence 1 in 1,000 to 1 in 2,500 live births per year		Birth prevalence 267–669 per year  (669,000 UK live births – 2006) Birth	26-64 (64191 live births – 2007)
Barth syndrome	Birth prevalence 1 in 300,000 to 1 in 400,000 per year USA		prevalence 152-203 per year  Birth	Birth prevalence 15-19 per year (9.6% of UK births)
Dystrophinopathies		Birth prevalence 4 in 18,500 live male births	prevalence 74 per year  (342,000 UK live male births –2006) 184,669–	7 (32095 live male births – 2007)
Total (excludes FH)			258,298	17438- 23814

This estimate of prevalence for DCM is based on sparse literature and is thought to be an underestimate (Perry Elliott, personal communication). The figures shown

are calculated on the assumption that approximately 25% of DCM is familial in nature.

The following conditions are not listed due to lack of identified prevalence data in the published literature: familial atrial fibrillation, short QT syndrome, progressive familial heart block, Loeys-Dietz syndrome, thoracic aneurysms and aortic dissections, mitochondrial cardiomyopathy and Emery-Dreifuss muscular dystrophy, bicuspid aortic valve.

### **Evidence base:**

The following national policy documents are *directly* relevant to ICC services:

- The National Service Framework for Coronary Heart Disease, Chapter Eight: Arrhythmias and Sudden Cardiac Death (2005)
- 'Heart to Heart'; Inherited Cardiovascular Conditions: A Needs Assessment and Service Review, The PHG Foundation (2009)
- The National Service Framework for Children, Young People and Maternity Services (2004)
- 'Transition: moving on well' – Good practice guide. Department of Health (2009)
- ICC services should also follow the National guidance found within the National Service Framework chapter for arrhythmias and sudden cardiac death: Implementation documentsii.
- The Association for Inherited Cardiac Conditions (AICC) is the UK national professional body for those involved in ICC management. The AICC developed with support from the Department of Health and provides expert guidance and care pathways for ICC management.

The following guidelines will be used where appropriate:

- Clinical indications for genetic testing in Familial Sudden Cardiac Death Syndromes: an HUK position statement (Garratt 2008)
- The American College of Cardiology/American Heart Association/European Society of Cardiology 2006 guidelines for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death (Zipes DP 2006)
- Guidelines produced by the Task Force of the European Society for Cardiology on Sudden Cardiac Death (Priori 2001)
- The Ghent Criteria for the diagnosis of Marfan syndrome (De Paepe 1996)
- Guidelines supporting Chapter 8 on the Department of Health website (includes implementation documents on Wolff-Parkinson White syndrome, Acquired LQT syndrome, Brugada syndrome, congenital LQT syndrome, HCM and ARVC) (Department of Health 2005)
- Guidance and care pathways from the Association for Inherited Cardiac Conditions (AICC) (<http://www.aicc-uk.co.uk/>)

## **2. Scope**

### **2.1 Aims and objectives of service**

#### **Aims**

The aim of ICC services is to improve the diagnosis, treatment and outcome of patients with inherited cardiac conditions. Improving outcomes in patients with ICC requires specialist clinical management to improve both life expectancy and quality of life.

#### **Objectives**

The service will deliver these aims by establishing defined models of care with robust shared/network care arrangements where appropriate to ensure:

- Timely diagnosis with appropriate counselling and psychological support to the patient and their family.
- Provision of high quality proactive treatment and care.
- A smooth and managed transition from children's care to adult care.

### **2.2 Service description/care pathway**

The ICC services provide care/treatment for infants, children, young people and adults. Cases of sudden death in infants less than one year old should be referred to the paediatric service.

The ICC service will need to investigate suspected index cases. Where an index case is identified, screening should be offered to first degree relatives, and cascaded to others as deemed necessary on the basis of risk.

The ICC service will function as a multi-disciplinary team (MDT). The MDT will have oversight of the whole service, which includes the following:

- Receiving referrals
- Provision of advice about possible referrals
- Diagnosis and assessment
- Treatment or advising on treatment
- Identification of family members at risk and recommending appropriate evaluation
- Discharge
- Prioritisation of services for patient, such as access to genetic testing, according to clinical need
- Input to specialist care provided by other specialities (for example ophthalmology, rheumatology, cardiac surgery)
- Input to long term monitoring, surveillance and care, including shared care arrangements with other hospitals and support provided by voluntary

organisations

- Providers will exercise their own judgement and experience, within the agreed parameters of any national commissioning policy, when recommending treatments for patients. The guiding principle will be that whatever treatment is recommended, it will be offered after discussion between the patient and clinician and be in the best interest of the patient.

### **Staff, training, qualifications and experience**

As stated in the National Blueprint for SCD and ICC services (endorsed by the DH 2007), the core team of staff will include:

- Consultant Cardiologists and Consultant Paediatric Cardiologists with specific expertise and experience in the management of ICCs.
- Consultant Clinical Geneticists and Genetic Counsellors to provide pre- and post- test counselling and to co-ordinate DNA testing, aid in genetic data interpretation and cascade testing of at-risk family members.
- ICC Nurse specialists with training in counselling, and in the evaluation and management of adults and children with inherited cardiovascular conditions.
- Echo technicians with specific training and expertise in the evaluation of inherited cardiovascular conditions.
- Echo technicians with specific training and expertise in the evaluation of inherited cardiovascular conditions in children.

All of these specialists may not necessarily be present on the same site or available at the same visit, but will be accessible to the ICC service

ICC services will ensure that:

- The MDT meets on a defined and regular basis. This will be no less than once a month.
- The MDT includes input from cardiology, genetics and, when necessary, from paediatric cardiology.
- The outcomes of MDT discussions are clearly recorded in the case notes of the relevant patients.
- The MDT oversees transition from the paediatric to the adult components of the service.
- There is robust evidence of clinical audit.
- Locally and regionally agreed clinical protocols are followed.

### **Diagnostic Facilities**

Centres providing the service will have access to the following:

- Dedicated echo service.
- Access to and experience with cardiac magnetic resonance imaging in cardiomyopathy.
- Exercise testing (risk stratification and diagnosis).
- Ambulatory electrocardiographic monitoring.
- Signal averaged electrocardiograms.

- Facilities for non-invasive or minimally invasive electrophysiology investigation, e.g. Ajmaline testing.
- Genetic testing, including specific blood tests which relate directly to ICCs (such as alpha-galactosidase testing for Fabry disease)

See Annex 1a for a generic patient pathway

### **Discharge criteria**

The following groups of patients may be considered for discharge from the ICC service:

- Patients at no risk.
- Patients at low risk for whom the risk unlikely to increase and who are unlikely to benefit from intervention. This group may be discharged to a GP or secondary care cardiologist for continued monitoring.
- Adult patients for whom no further intervention is required at this time, e.g. (i) HCM with no associated risk factors and symptomatically well, or (ii) Marfan syndrome with normal aorta and mitral valves. This group may be discharged to a secondary care cardiologist for continued monitoring.

At risk children will be followed up until they reach adulthood, when assessment of the most appropriate future care will be made.

Patients with a progressive condition who could potentially benefit from treatment at a later date will not normally be discharged.

Patients will usually be discharged to the care of their General Practitioner or local Hospital Cardiologist.

### **General Paediatric care**

*When treating children, the Service will additionally follow the standards and criteria outlined in the Specification for Children's Services (attached as Annex 1 to this Specification)*

### **2.3 Population covered**

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

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



The ICC services provide care/treatment for infants, children, young people and adults. Cases of sudden death in infants less than one year old will be referred to the paediatric service.

Anyone with any of the conditions listed in section 1.1, or with a first degree relative with one of these conditions, will be able to access the ICC service. This will include the next of kin in cases of sudden cardiac death.

The service will be delivered at times that meet individual's needs. The service will be accessible and in an appropriate setting. The service will be delivered in a format and style, which recognises the needs and preferences of all patients, which include:

- different cultural or faith groups
- physical/learning disabilities
- ethnic minorities

The provider will have a clear equality and diversity policy (we recognise that the provider will need to treat groups differently, so a clear policy on how this will be done sensitively and reflecting the evidence base will be required).

The provider will make information available to patients and the public on their services, provide patients with suitable and accessible information on the care and treatment they receive and, where appropriate, inform patients on what to expect during treatment, care and after-care.

The onus upon services to monitor the ethnic origins of their patients might also have important clinical consequences, given emerging epidemiological evidence suggesting that for some conditions, such as cardiomyopathy, there is an increased predisposition for the development of disease in certain ethnic groups

## **2.4 Any acceptance and exclusion criteria**

### **Acceptance Criteria**

The service will accept inward referrals from secondary and tertiary care for patients with suspected Inherited Cardiac Conditions.

### **Referral Criteria**

Anyone with any of the conditions listed in section 1.1, or with a first degree relative with one of these conditions, shall be able to access the ICC service. This will include the first degree relatives in cases of sudden cardiac death.

All patients with an ICC, or potentially at risk of having an ICC, should be offered assessment in a specialist ICC service, or advice taken from that service. They should not be managed by non-ICC cardiologists, geneticists or other specialties

alone, without comprehensive ICC service involvement.

Patients with an ICC will have the initial diagnosis made or confirmed within a specialist ICC service. The ICC service will determine the initial management plan, assess the pedigree, set up genetic testing and arrange family evaluation and cascade screening where appropriate.

The follow up plan may involve the ICC service or may be devolved to local hospital cardiologist or primary care depending on patient risk and complexity.

The ICC service will need to investigate suspected index cases. Where an index case is identified, screening shall be offered to first degree relatives, and cascaded to others as deemed necessary on the basis of risk.

ICC services will be embedded in secondary and primary care services that are able to recognise individuals at risk, refer appropriately, support patients and families in interpreting and acting on specialist advice, and provide a partnership for shared care and follow-up. This requires that all professionals in cardiology, primary care, paediatrics and any of the secondary and tertiary specialties that interface with inherited cardiac disease have a necessary basic level of understanding and competence.

## **2.5 Interdependencies with other services**

The ICC service will be available and accessible to all its catchment population. Whilst it is usually necessary that it should be sited geographically in a major tertiary centre (often where there is a regional genetics service) the service will find means to reach out to its feeder health and other services to ensure that individuals with ICCs are diagnosed and managed effectively.

Where outreach services are agreed, assurance will be given that the same quality of service can be delivered and that the designated service retains responsibility for the overall delivery of the service including clinical governance. There will be clear arrangements to define responsibility for service quality, clinical governance (including, but not restricted to, clear arrangements for dealing with complaints, serious untoward incidents and clinical indemnity).

### **Co-located services**

Certain elements of the ICC service need to be co-located including the Consultant ICC Cardiologist, ICC Nurse Specialists and basic non-invasive testing facilities such as Electrocardiogram (ECG), holter monitoring, exercise stress testing and echocardiography. Ideally the cardiology elements of the ICC service should also be co-located with those interdependent cardiology specialties given below. The genetic elements of the ICC service, Consultant Clinical Geneticist and Genetic Counsellors, should also be co-located.

It is accepted that ICC Cardiology specialist services and Genetic Services may not necessarily be physically housed in the same building or on the same site, but they should function as a co-located service. Due to the arrangement of paediatric services across the UK, paediatric ICC cardiology or metabolic disease specialties may also not be physically present on the same site, but the ICC service should have established pathways of access to these services.

Specialist cardiac pathology should ideally be co-located with the main ICC centre. Where this is not possible, the link should be well-established such that the two elements of the service are functionally co-located.

Molecular genetic testing laboratories may be remote from the ICC service.

### **Interdependent services**

In addition to the core team, ICC service should have close links with electrophysiology, diagnostic angiography, interventional cardiology, cardiothoracic surgery and cardiovascular imaging departments.

### **Related services**

Individuals and families accessing the ICC service shall also have access to dedicated bereavement counselling services, through either their regional ICC service or the national support groups

## **3. Applicable Service Standards**

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

#### **Core Standards**

The core team of staff will include:

- Consultant Cardiologists and Consultant Paediatric Cardiologists with specific expertise and experience in the management of ICCs.
- Consultant Clinical Geneticists and Genetic Counsellors to provide pre- and post- test counselling and to co-ordinate DNA testing, aid in genetic data interpretation and cascade testing of at-risk family members.
- Nurse specialists with training in counselling, and in the evaluation and management of adults and children with inherited cardiovascular conditions.
- Echo technicians with specific training and expertise in the evaluation of inherited cardiovascular conditions.
- Echo technicians with specific training and expertise in the evaluation of inherited cardiovascular conditions in children.

#### **ICC services will ensure that:**

- The MDT meets on a defined and regular basis. This will be no less than once a

month.

- There is robust evidence of clinical audit.

### **Diagnostic Facilities**

Centres providing the service will have access to the following:

- Dedicated echo service.
- Access to and experience with cardiac magnetic resonance imaging in cardiomyopathy.
- Exercise testing (risk stratification and diagnosis).
- Ambulatory electrocardiographic monitoring.
- Signal averaged electrocardiograms.
- Facilities for non-invasive or minimally invasive electrophysiology investigation, e.g. Ajmaline testing.

### **Recommended Standards**

Specialist ICC services should follow recommendations for standards of care, best practice, care pathways, treatment algorithms, data collection and audit, as produced by the Association for Inherited Cardiac Conditions (AICC), National Institute for Health and Clinical Excellence (NICE) or other recognised national professional bodies.

### **Audit**

There will be a robust internal monitoring system evaluating outcomes to allow the early identification and pre-emption of any adverse outcomes. There will be regular audit meetings and discussions of mortality and serious adverse events.

The service shall contribute to national service reviews and audits, including any new processes introduced by the Association for Inherited Cardiac Conditions

### **Guidelines and guidance**

- Compliance with requirements of NICE guidance should be in line with legal requirements  
[http://www.nice.org.uk/media/8BD/2B/Legal\\_context\\_nice\\_guidance.pdf](http://www.nice.org.uk/media/8BD/2B/Legal_context_nice_guidance.pdf)
- ICC services shall also adhere to any other relevant National targets (including Performance Targets within the NHS Operating Framework)

The following national policy documents are *directly* relevant to ICC services:

- The National Service Framework for Coronary Heart Disease, Chapter Eight: Arrhythmias and Sudden Cardiac Death (2005)
- 'Heart to Heart'; Inherited Cardiovascular Conditions: A Needs Assessment and Service Review, The PHG Foundation (2009)
- The National Service Framework for Children, Young People and Maternity Services (2004)
- 'Transition: moving on well' – Good practice guide. Department of Health

(2009)

ICC services shall also follow the National guidance found within the National Service Framework chapter for arrhythmias and sudden cardiac death:

Implementation documents<sup>iii</sup> and recommendations and care pathways from the Association for Inherited Cardiac Conditions. These documents cover:

- Initial treatment
- Ongoing treatment
- Sudden Cardiac Death
- Patient support

The following guidelines are identified in the PHG Foundation Report and shall be used where possible:

- Clinical indications for genetic testing in Familial Sudden Cardiac Death Syndromes: an HUK position statement (Garratt 2008)
- The American College of Cardiology/American Heart Association/European Society of Cardiology 2006 guidelines for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death (Zipes DP 2006)
- Guidelines produced by the Task Force of the European Society for Cardiology on Sudden Cardiac Death (Priori 2001)
- The Ghent Criteria for the diagnosis of Marfan syndrome (De Paepe 1996)
- Guidelines supporting Chapter 8 on the Department of Health website (includes implementation documents on Wolff-Parkinson White syndrome, Acquired LQT syndrome, Brugada syndrome, congenital LQT syndrome, HCM and ARVC) (Department of Health 2005)

#### **4. Key Service Outcomes**

Service provision is focused on achieving optimum clinical management.

For many ICCs, careful clinical management will reduce risk of worsening clinical conditions or death. For arrhythmia syndromes (such as LQT) and HCM, treatments can include a variety of drugs, lifestyle advice to avoid triggering events, fitting of implantable defibrillator devices or, in some cases, surgical options.

Genetic testing can aid clinical management by enabling more accurate diagnosis and risk assessment, and in some cases guiding the choice of treatment. Importantly, identification of a mutation in an affected person enables their relatives to be offered targeted testing for the same mutation. Those who carry the mutation can then be assessed and offered appropriate surveillance and/or treatment, while those who do not carry the mutation can be spared further investigation. This approach is known as cascade testing.

## References

- Department of Health (2004) National Service Framework for Children, Young People and Maternity Services:
- Department of Health (2005) National Service Framework for Coronary Heart Disease; Chapter Eight: Arrhythmias and Sudden Cardiac Death
- Department of Health (2009) Transition: moving on well.
- Burton H, Alberg C, Stewart A. PHG Foundation (2009) Heart to Heart: Inherited Cardiovascular Conditions Services - A Needs Assessment and Service Review. A. ISBN 978-1-907198-01
- McKenna W et al. (2007) Proposal for the establishment of inherited cardiovascular conditions centres
- Services for Ehlers-Danlos Syndrome are provided by two national services located in London and Sheffield

## **ANNEX 1 TO SERVICE SPECIFICATION:**

### **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 (HSC) 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 (DOH) & 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 – DOH

## Imaging

All services will be supported by a 3 tier imaging network ('Delivering quality imaging services for children' DOH 13732 March2010). 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 (CPD)
- 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.<sup>1</sup> 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) training<sup>2</sup> and should maintain the competencies so acquired<sup>3\*</sup>.

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 an 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 on the Provision of Anaesthetic Services (GPAS) Paediatric anaesthetic services. Royal College of Anaesthetists (RCoA) 2010  
[www.rcoa.ac.uk](http://www.rcoa.ac.uk)
2. Certificate of Completion of Training (CCT) in Anaesthesia 2010
3. 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 persons 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 dietitians, 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:

- Ensures the medicines given are appropriate and person-centred by taking account of their age, weight and any learning disability
- Ensuring that staff handling medicines have the competency and skills needed for children and young people's medicines management
- Ensures that wherever possible, age specific information is available for people about the medicines they are taking, including the risks, including information about the use of unlicensed medicine in paediatrics.

Many children with long term illnesses have a learning or physical disability.

Providers should ensure 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 Publications, 2006, London

## Annex 1a

### Patient Pathway for Children and Adults who may have an Inherited Cardiac Condition (ICC)

This is a generic care pathway designed for the purposes of this service specification. It shows how patients should move through the ICC service.

