

Adult Congenital Heart Disease

A commissioning guide for services for young people and Grown Ups with Congenital Heart Disease (GUCH)

DH INFORMATION READER BOX

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Foreword

There are around 135,000 young people and adults currently living in England with congenital heart disease. This guide sets out how the National Health Service (NHS) can best meet their needs for high quality, responsive and cohesive services.

The Bristol Inquiry into children's heart surgery in July 2001 signalled the need for fundamental change to the way the NHS cares for children with congenital heart disease. The Independent Review of NHS Paediatric and Congenital Cardiac Services commissioned by the Department of Health as part of its response to the Inquiry set standards and made a series of recommendations to ensure the safe organisation of the paediatric and congenital cardiac service.

Many of those recommendations also apply to services for adults with congenital heart disease. The Review identified transition to adult services as perhaps the main outstanding area where services needed to be improved.

At a time when the number of adults with congenital heart disease has overtaken the number of children with congenital heart disease, this guide has been produced in response to those recommendations. It is specifically for adult services and for 'transition to adult' services, taking into account the Review's recommendations and other key guidelines relevant to adult services.

It has been compiled by an External Reference Group of experts in the field from the voluntary sector and the NHS and is based on the views of a wider stakeholder group comprising patients, families, carers and NHS staff. It will assist the NHS in planning and developing services, and will inform patients and their families what they can expect of the NHS.

I would like to thank all the members of the External Reference Group and the stakeholder group for their contributions.



Rosie Winterton

Minister of State for Health Services

Introduction

The National Service Framework for Coronary Heart Disease was published in March 2000 and considerable progress has been made in prevention, treatment, quality of care and access to care in this important area of heart disease. One topic that the Framework did not address was that of congenital heart disease.

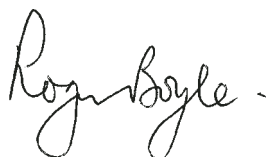
There have been major advances in the care of congenital heart disease over the last two decades so that there are now many more neonates and children surviving into adult life. But we know that up to one third of those with more complex problems will require further intervention later in life.

In the past, services for adults with congenital heart disease have been somewhat fragmented and the experiences of families as they pass through the transition from paediatric to adult services have been less than ideal.

The patient groups are quite clear that they wish to see better organisation of services, including a concentration of expertise, so that clinical services can properly meet the complex needs of such patients. At the same time, attention needs to be paid to the detail so that generic care such as anticoagulant control can be delivered close to home.

This document has been drawn up in conjunction with patients, patient groups and experts in order to help commissioners of care drive improvements for this group of patients, many of whom have spent their entire lives in close contact with the NHS and have strong views about how their care should be delivered.

As we continue to develop a system of commissioning that delivers a patient-centred service, we need to ensure that these voices are heard. This document is intended to help commissioners with this task.



Professor Roger Boyle
National Director for Heart Disease

Executive summary

The purpose of this guide is to provide advice to commissioners of NHS services and cardiac networks and to inform patients' expectations of NHS services.

It covers services for people with congenital heart disease age 16 and over with all types of congenital conditions.

The strategic context section lists other guidance and publications such as the National Service Framework for Long-term Conditions, which are relevant to planning congenital heart disease services including recent reports specifically about services for adults with congenital heart disease, which have informed the External Reference Group in producing this guide.

The reforms of the NHS and other changes such as the European Working Time Directive will impact upon services for adults with congenital heart disease. These are set out in the section on 'Drivers for change'. Of particular note is the growth in population of adults with congenital heart disease, which has highlighted the need for a more cohesive NHS service response for adults.

The External Reference Group suggests a direction of travel to achieve more cohesive high quality services involving the concentration of expertise in a small number of specialist centres and raising awareness of congenital heart disease in other parts of the NHS. A distinction is drawn between specialist centres and more local centres. Local centres would be able to provide shared care under protocols agreed with a specialist centre. A possible service model is illustrated. The model also shows dedicated transition services between paediatric and adult services.

Suggestions are made to assist local planning such as defining the referral route for patients to access appropriate NHS expertise and clarifying the facilities, skills and knowledge available at each centre. Indicators of high quality care are provided covering:

- Transition services
- Access to an expert
- A multi disciplinary approach (including primary care, oral health and obstetric care)
- Special needs

The Department of Health support for implementing this guide includes work on clinical audit, clarifying guidelines on antibiotic prophylaxis and continuing to work with patient groups and NHS professionals to oversee progress.

The final two sections of the guide form a resource pack comprising:

- Where to find key documents, which have informed the work on the guide;
- Guidance on estimating the population of adults with congenital heart disease;
- Suggested facilities and skills needed at specialist centres and local centres;
- Good practice in transition services;
- References to documents to assist workforce planning and training including the medical competencies for cardiologists at specialist and local centres;
- Suggested skills and knowledge areas for nurse specialists.

Purpose and scope of this guide

Purpose

1. The purposes of the guide are:
 - To provide advice, guidance and support for Primary Care Trusts (PCTs) commissioners, local specialised commissioning groups (LSCGs), Strategic Health Authorities, cardiac networks and paediatric services in the monitoring, planning and development of services.
 - To inform patients' and their families' expectations of NHS services by providing indicators of high quality care.
2. Local services will need to agree which aspects of the Guide are relevant to local circumstances and to their patients' needs and adapt them accordingly.
3. The focus of the Guide is on organising NHS services to deliver most efficiently to improve quality of care and patient experience of services. It is about using resources to best effect.

Scope

4. The guide applies both to services for young people and to services for adults with congenital heart disease (also called Grown Ups with Congenital Heart Disease or GUCH). The adult population is taken to be age 16 and over in this document. (Children's services are addressed in the Paediatric and Congenital Cardiac Services Review Report). Congenital heart disease describes a range of heart conditions resulting from an abnormality of the heart structure or function that is present at birth. Most conditions are a result of the heart or its valves and vessels not being properly formed. There may also be holes between the chambers of the heart. There are many different types ranging from simple defects with minor consequences to complex defects with serious outcomes. Some people will require little or no follow up after an appointment with a specialist but others will need to be followed up throughout their lives and may have episodes of illness requiring hospital care. People with congenital heart disease have a greater risk of becoming ill. Congenital heart disease also affects employment, further education, relationships and leisure pursuits.

Strategic context

The following list identifies existing guidance which is relevant to planning and commissioning of services for adults with congenital heart disease:

5. *The NHS Improvement Plan* published in June 2004 sets out the vision for the NHS and the key commitments that the NHS is expected to deliver up to 2008. The focus is on more personalised care including more choice for patients, more support in the community and at home and an increasing emphasis on quality and safety of services.
6. *National Standards, Local Action, Health and Social Care Standards and Planning Framework 2005/06-2007/08* sets the standards and targets which will drive improvement in quality and clarifies expectations of the way that the NHS plans and delivers services.
7. *The National Service Framework (NSF) for Long-term Conditions* published in March 2005 sets out quality requirements and good practice to improve services for people with long-term conditions. The focus is on neurological conditions but PCTs are encouraged to use the NSF in planning any service developments for people with other long-term conditions such as congenital heart disease.
8. *Our health, our care, our say: a new direction for community services a white paper* published in January 2006 includes proposals to improve support for people with long term needs and to give people a stronger voice in improving services.
9. More specifically, in relation to services for adults with congenital heart disease, the *Paediatric and Congenital Cardiac Services Review Report* published in November 2002 sets out evidence-based standards for all paediatric and congenital cardiac services which, where appropriate, should also apply to services for young people and adults.

10. In September 2004, the *National Service Framework for Children, Young People and Maternity Services* included a standard on the needs of young people growing up into adulthood. More recently a good practice guide called *Transition: getting it right for young people* was published in March 2006 showing how the handover from children's to adult services can be planned and managed.
11. The Report of the British Cardiac Society Working Party on *Grown-up Congenital Heart Disease (GUCH): current needs and provision of service for adolescents and adults with congenital heart disease in the UK (September 2002)* makes recommendations on the organisation of national medical care, training of specialists and education of doctors and nurses to address the needs and problems of congenital heart disease patients . This report, together with the report of the European Society of Cardiology's Task Force on *Management of Grown Up Congenital Heart Disease (June 2003)*, has informed the work of the External Reference Group in preparing this Guide.

The Department of Health Publications mentioned above can be found at:
<http://www.dh.gov.uk/PublicationsAndStatistics/Publications/fs/en>

Drivers for change

12. The NHS is in the process of implementing financial and organisational reforms. In 2006/7 the pace of reform is being stepped up. Priorities for 2006/7 are set out in *The NHS in England: the operating framework for 2006/7* published in January 2006. Of relevance to services for adults with congenital heart disease are:
 - Extension of Payment by Results: Payment by Results (PbR) is a transparent, rules based system, which is transforming the way key NHS services are funded by paying hospitals according to the number and complexity of cases treated. PbR is a prospective payment system that applies tariffs based on the average costs of NHS services as reported by the NHS organisations themselves. Tariff setting is itself a clear and transparent process in that the national prices can be traced back to the reference costs data supplied by the NHS. The tariff is structured around clinical casemix measures known as Healthcare Resource Groups (HRGs) which represent clinically coherent groups of diagnoses and treatments that consume similar levels of healthcare resource. PbR rewards efficiency and supports innovation by allowing managers and clinicians the freedom to retain any financial surplus for reinvestment in services. PbR is necessary to support patient choice because it allows money to follow the patient and therefore rewards providers that attract people by responding to their need and preferences.
 - Extension of patient's choice: From January 2006, patients needing elective treatment are being offered a choice of appointment and at least four providers once their GP has decided that a referral is required across a number of specialities, including cardiology. For people with congenital heart disease, in most cases, it is likely to be more important to receive 'expert' care than the quickest care and for appropriate cardiac congenital expertise to be readily accessible when needing treatment for non-cardiac conditions.

13. Waiting list target: another driver which will impact on services for adults with congenital heart disease is that by December 2008, no-one waits more than 18 weeks from GP referral to hospital treatment. This applies to acute elective hospital care and will encompass all diagnostic procedures and tests. PCTs will be planning for reductions in diagnostic waits. These improvements in diagnostics will benefit adults with congenital heart disease. There are interim targets that no one should wait for more than six months for inpatient elective care and more than three months for an outpatients appointment (those associated with acute elective care) from December 2005.

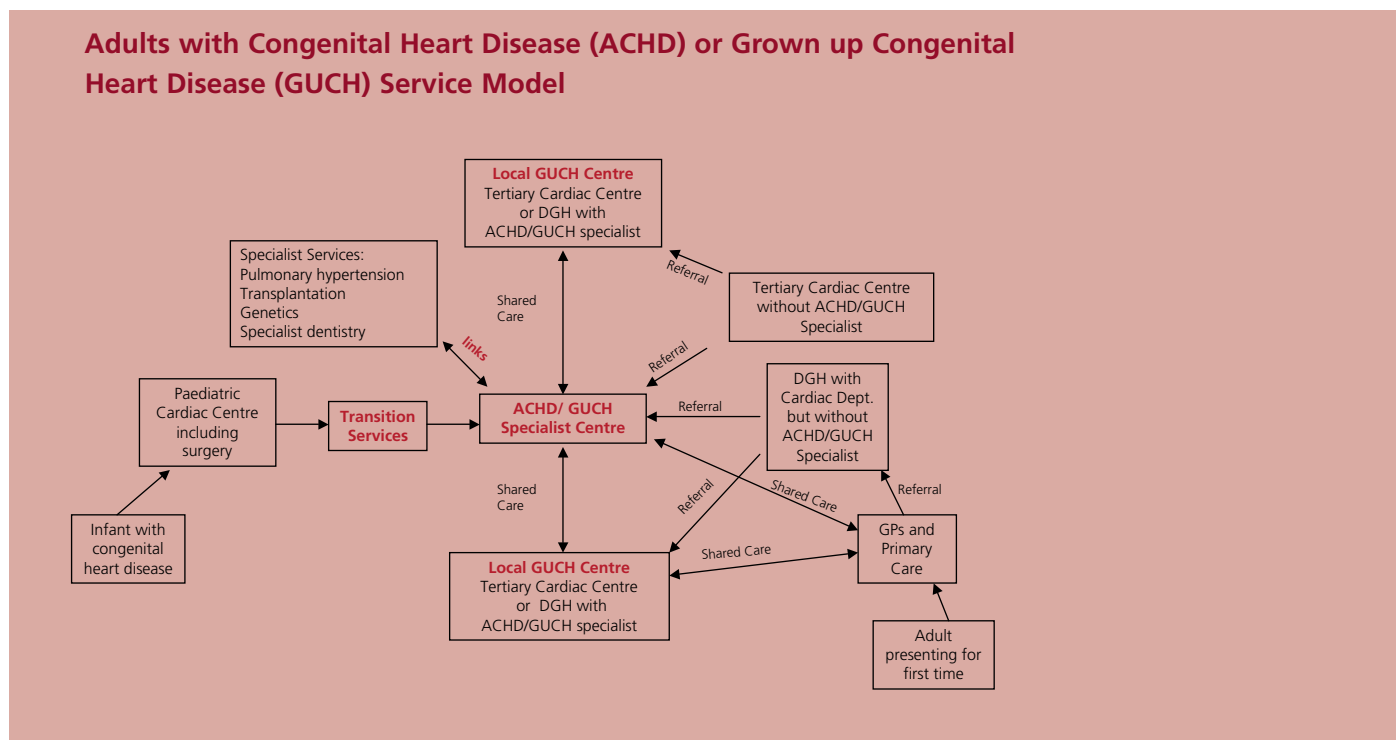
Other drivers relevant to services for adults with congenital heart disease are:

14. Population growth: The success of paediatric cardiology and surgery has led to a significant increase in the population of adults with congenital heart disease. The 18-year survival rate for complex defects was 10% for those born before 1960 but is 50% for those born in the 1980s. Adults with congenital heart disease now outnumber children with this condition and the adult population will continue to rise (see paragraph 53). A high proportion of adults with learning disabilities have congenital heart disease and the number of learning disabled adults is also increasing (see paragraph 54)
15. This 'new' and growing population does not sit easily within the traditional divisions of training and practice for adult and paediatric cardiology suggesting the need for a different service response. Patients need a more cohesive service, which recognises that congenital heart disease is a life long condition, which affects quality of life.
16. Changes to medical training: The Government launched its 'Modernising Medical Careers' strategy in February 2005. This proposes reforms of postgraduate medical training including development of more effective team work, multi disciplinary working and working in different settings to respond to changing models of care. All of these will be important for delivery of services for adults with congenital heart disease.
17. For services for adults in congenital heart disease, there is a need both to secure and build a specialist workforce for a growing population and, in addition, meet the requirements of the European Working Time Directive (EWTD). The UK Working Time Regulations enacted as a result of the EWTD lays down minimum requirements for employers in relation to working hours, rest periods and annual leave. The most significant of these is the maximum 48-hour working week which is currently being phased in for doctors in training in the UK and needs to be achieved by 2009. This may mean developing new ways of service delivery.
18. Clinical audit: The Healthcare Commission, an independent body, which regulates and promotes improvement in the quality of healthcare and public health, plans to increase attention to quality and effectiveness of treatment. This is focusing attention on the importance of developing better data systems to enable clinical audit and research to inform future treatment strategies for both paediatric and adult congenital heart disease services.

The direction of travel

19. The External Reference Group agreed a preferred direction of travel for services for adults with congenital heart disease or GUCH services. The aim is to create a more cohesive high quality service where expertise is concentrated in a small number of specialist centres and at the same time to raise awareness of congenital heart disease in other parts of the NHS, including primary care. A shared care model (where care is shared between a local centre, a specialist centre and the patient's GP practice) offers opportunities for better cohesion and improved access to services whilst retaining the expert overview of service delivery.

Figure One below shows a possible service model to assist with planning and implementation of improvements:



20. This model shows dedicated transition services (explained in paragraph 60) to facilitate the entry of young people into adult services. It also uses the term “local GUCH centres” to define those tertiary cardiac centres (see glossary) or District General Hospitals (DGHs) where there is a cardiologist with expertise in adults with congenital heart disease. Local GUCH centres would provide shared care under protocols agreed with the specialist centre for the majority of patients. Some people may present as adults to their GP with a congenital condition which has been previously undiagnosed or, possibly, after losing touch with services provided when they were children. The model shows a referral route in these cases.

Implementation of service improvements

21. In following this direction of travel, commissioners, Strategic Health Authorities and cardiac networks will need to come to a local agreement on relative priorities and decide on the pace of change in implementing service improvements. The following points are offered for consideration and to assist local planning.

Suggested points for cardiac networks, commissioners and Strategic Health Authorities to consider:

- Is there a defined referral route for young people and adults presenting with possible congenital heart disease for the first time, which makes it clear that they are always referred to identified specialist centres or identified local GUCH centres for assessment and treatment including for non-cardiac conditions?
- Is the role of specialist centres, local GUCH centres and other NHS services clearly understood within the local NHS and by patients with congenital heart disease?
- Is the network aware of the facilities, skills and knowledge required at specialist centres and local GUCH centres? Paragraphs 56 and 57 give an example of these for specialist centres and local GUCH centres based on current European guidelines and on advice from the External Reference Group.
- Do commissioners know the local GUCH population and have any projections been done to establish the likely population of patients with simple, medium and complex conditions up to 2010? Paragraph 53 provides estimates by Strategic Health Authority (SHA) for discussion with specialist adult and paediatric centres to assist in planning assumptions.
- Has any review of services been carried out?
- The following indicators of high quality care are offered to assist local service review and inform local development.

Local indicators of high quality care

22. Commissioners may find it helpful to adopt all or some of these indicators as guides to good practice or developmental goals in local consideration and planning of services. While they are not national requirements, they represent a consensus among patients, parents, health professionals and service managers as to what constitutes good practice and should be helpful in enabling the local assessment and improvement of services.

Transition services

23. Indicators of high quality care

- Every paediatric cardiac unit has a transition arrangement with an adult specialist unit, which is separate from paediatrics and services for adults and, ideally, has dedicated inpatient accommodation available within adult services.
- All entrants to adult services receive a detailed written care plan, identifying the specialist or local GUCH centre where applicable and stating the follow up arrangements, responsibilities and intervals. Plans are copied to the local GUCH centre and the patient's General Practitioner (GP). The plan forms the basis of the patient held record and copies of future consultant/ GP letters are sent to the patient for adding to this record.
- A nurse specialist is available as the main contact point for each patient in transition.
- Transition does not automatically occur at age 16 but takes into account the needs and wishes of the young person. Transition is normally completed by age 18.
- Local outpatient booking systems allow for long-term follow up appointments at appropriate intervals which could be every five years or even ten years.

Access to an expert

24. Indicators of high quality care – care planning

- All adults with congenital heart disease **whatever the level of complexity** are seen by an ‘expert’ from a specialist centre at least once and receive a written care plan. The majority of patients may expect to receive their care under shared care arrangements between a specialist centre and a local GUCH centre. Certain conditions will always require a specialist input but others will vary throughout a person’s life and may become complex for brief periods.

25. Indicators of high quality care – specialist centres

- Specialist centres have shared care protocols with local GUCH centres and GPs. This may include outreach services.
- Specialist centres maintain close links with paediatric cardiac centres.
- Specialist centres have established links with other relevant specialist services such as genetics, transplantation and the National Pulmonary Hypertension Service.
- Specialist centres provide a 24-hour point of reference for colleagues in local GUCH centres, tertiary centres, DGHs and primary care including general dental practitioners and community dental clinics.
- Specialist centres provide teaching and training including training for technicians and also post-graduate education.
- Specialist centres work collaboratively with each other and local GUCH centres to ensure national audit.
- Specialist centres are committed to research and development and provide an environment where research can be carried out.

26. Indicators of high quality care – local GUCH centres

- Local GUCH centres are able to provide shared care under protocols agreed with a specialist centre to avoid the need for patients to travel to a specialist centre for follow up where this is a long distance.
- If they prefer, patients may choose to attend a different centre from their local GUCH centre. In some cases the specialist centre will be the most local centre to where the patient lives.
- Local GUCH centres have a nominated cardiologist with skills in adult congenital heart disease (see paragraph 64).
- Local GUCH centres have a clear understanding of which conditions they can treat and clear protocols about what and when to refer to a specialist centre.

A multi disciplinary approach

27. Indicators of high quality care

- All young people and adults with congenital heart disease receive care delivered by a multi disciplinary, multi speciality team. Within the NHS, the implications of their cardiac condition are taken into account within other specialties and there is expertise available in other relevant specialties such as obstetrics, anaesthetics and dentistry.
- Patients have access to ongoing support and advice on living with their condition and in particular follow up after a period of acute care. They also have ready access to information on understanding their condition and on accessing services which is appropriate to their level of understanding.
- For every congenital heart disease patient, GPs and the identified local GUCH centre receive communications from specialist centres on diagnosis, prognosis, the care plan and information to support the patient.
- Patients receive ongoing support and advice from a specialist GUCH nurse.
- Specialist centres provide a 24-hour point of reference for patients on any aspect of care including, for example, contraception, pregnancy and dental care.
- Specialist centres and local GUCH centres promote patient support groups such as the GUCH Patients Association and establish local support groups including support for carers.

- Patients are encouraged to monitor themselves and are provided with guidelines for doing this. Trusts may consider providing International Normalised Ratio (INR) testing devices for pro thrombin time for use by patients to minimise attendance at hospital (see glossary).

28. Indicators of high quality care – primary care

- GPs and other primary care staff are part of shared care arrangements with specialist centres and local GUCH centres. They manage routine cardiac medication and monitor anticoagulation and blood chemistry.
- For patients with congenital heart disease, GPs and primary care staff are clear about referral routes, services and sources of advice available including patient support groups.
- GPs understand their patients' diagnoses and are able to communicate this and to educate patients and their families and carers.
- GPs are sufficiently knowledgeable to know which other treatments for non- cardiac conditions will affect the patient's heart condition.
- GPs know when to refer for specialist advice (for example, for contraception) and where to refer in an emergency.
- GPs know about the risks of endocarditis for GUCH patients associated with dental and surgical procedures and body piercing and understand the requirements for prophylaxis.

29. Indicators of high quality care – oral healthcare

- Dental surgeons are essential members of the multi disciplinary team at a specialist centre and are involved at an early stage in treatment planning and overall care as well as giving oral hygiene advice and advising on all other preventive measures.
- There is a joint approach with a multi disciplinary clinic involving cardiologists, dental surgeons, dental hygienist/therapist and dieticians. If intravenous antibiotics are not required, ongoing care may be undertaken by a general dental practitioner or at a community dental clinic.
- Specialist centres have access to a dedicated dental team (dentist, dental nurse, hygienist/therapist), providing acute care for inpatients and a screening service for patients undergoing surgery.
- Outpatients attend dental clinics on a regular basis for routine screening and reinforcement of oral hygiene.

- Specialist centres have lists of dental services where patients can obtain 'shared care' and information sheets for patients on dental management. Information sheets may be passed on by the patient to their local dentist.
- General Dental practitioners and community dentists have an understanding of the need for antibiotics and of anticoagulation issues and risks with sedation.

30. Indicators of high quality care –obstetric care

- Pregnant patients with complex cardiac conditions are managed at a specialist centre.
- Obstetric care for pregnant patients with simple conditions may be managed under shared care arrangements at a local GUCH centre.
- Specialist centres arrange joint clinics between the cardiologist and specialist obstetrician.
- Fetal medicine specialists and fetal echo services are accessible at the specialist centre.
- Pregnant patients with simple cardiac conditions attending local GUCH centres under shared care arrangements are referred to the specialist centre for fetal echo.

Special needs

31. Indicators of high quality care

- People with learning disabilities and other special needs have the same access and choices of services and their special needs are provided for.
- Patients with special needs requiring dentistry are managed in a specialist centre. (A significant number of patients with learning disability and congenital heart disease can only undergo dentistry under intravenous sedation or general anaesthesia. This can present problems in anaesthetic management and peri-operative care).
- Specialist centres have a clear and readily available source of specialist advice on learning disabilities including specifically advice on anaesthesia.
- Accommodation for families /carers is available for patients who need an inpatient stay.

Department of Health support for implementation

32. In October 2005, the Department of Health commenced a review of NHS commissioning arrangements for specialised services, which includes services for adults with congenital heart disease. The review is looking for ways of improving commissioning arrangements so that there is greater consistency and that the arrangements are fit for purpose and robust in the context of wider NHS reforms. The Review report is due in Spring 2006.
33. The Department is continuing to work with the Health and Social Care Information Centre and clinicians to ensure that the complexities of treatment of congenital heart disease are reflected in the next version of Healthcare Resource Groups from which new national tariffs will be devised. This is important so that specialist providers are able to charge for services at an appropriate level for the resources used in providing more specialist care as opposed to the standardised tariffs for general cardiac care.
34. The Department of Health will work with representatives from the British Congenital Cardiac Association, the Health and Social Care Information Centre and the Healthcare Commission to assess the work to expand the existing paediatric surgical/ catheter interventions clinical audit database to encompass all interventions for children, young people and adults. Specialist centres will need to ensure robust data collection, have a computer system with linked software and facilities for long-term electronic record storage.
35. Growth in the GUCH population and the implementation of the European Working Time Directive suggests that we need more training places for doctors and cardiologists with expertise in adult congenital heart disease. The Workforce Review Team will work with SHAs and Workforce Development Confederations to address this in planning assumptions.
36. The Department of Health will work with the British Congenital Cardiac Association to develop a list of potential research topics.

37. The Department of Health will consider proposals from appropriate dental bodies to develop protocols and best practice guidelines for the dental management of people with congenital heart disease, together with proposals for the use of these protocols and guidelines to facilitate both undergraduate and postgraduate training, subject to the availability of resources
38. The Department of Health is working with the National Institute for Health and Clinical Excellence (NICE) to achieve consensus over indications for antibiotic prophylaxis and those at risk of endocarditis. This will include consideration of the needs of those who wish to undergo body piercing or tattooing.
39. The Department of Health will continue to work with patient groups and professionals to oversee progress in improving service delivery for young people and adults with congenital heart disease.
40. Subject to consultation, the indicators of high quality care in this guide will inform the assessment of NHS services by the Healthcare Commission. The Department of Health will work with the Healthcare Commission to support the Commission's development of specific measurable indicators for services for adults with congenital heart disease.

Source documentation details

41. The following documents: have informed the development of this guide:

- European Society of Cardiology's guidelines on the 'Management of Grown Up Congenital Heart Disease' June 2003 (informed service model).
<http://www.escardio.org/knowledge/guidelines/>
- The British Cardiac Society Working Party on Grown-up congenital heart disease (GUCH): current needs and provision of service for adolescents and adults with congenital heart disease. September 2002 (informed service model).
http://heart.bmjournals.com/cgi/content/full/88/suppl_1/i1
- Paediatric and Congenital Cardiac Services Review Report November 2002 (recommended standards and identified issues for transition). This can be found at:
<http://www.dh.gov.uk/assetRoot/04/01/86/22/04018622.pdf>
- The National Service Framework for Children, Young People and Maternity Services September 2004: Standard 4 'Growing up into adulthood' and standard 8 'transition into adulthood for young people with complex needs' can be found at:
<http://www.dh.gov.uk/PolicyAndGuidance/HealthAndSocialCareTopics/ChildrenServices/ChildrenServicesInformation/fs/en>
- Transition: getting it right for young people can be found at:
http://www.dh.gov.uk/PublicationsAndStatistics/Publications/PublicationsPolicyAndGuidance/PublicationsPolicyAndGuidanceArticle/fs/en?CONTENT_ID=4132145&chk=09SMKE
- The 32nd Bethesda conference : Care of the Adult with Congenital Heart Disease JACC Vol 37, 2001 (population estimates model – see below) <http://www.acc.org/clinical/bethesda/beth32/tables.htm>
- The Canadian Cardiovascular Society's Consensus Conference update 2001 update(informed service model)
http://www.ccs.ca/download/consensus_conference/consensus_conference_archives/2001_adults_CHD.pdf

Further Information and Resources

Population needs assessment

42. The External Reference Group has advised the Department of Health that the numbers of adults with congenital heart disease is increasing significantly and possibly by as much as 10% per year (all conditions). However, it is difficult to produce accurate figures without detailed needs assessment work.
43. An example of population needs assessment work for adults with congenital heart disease can be found in North West England. North West Adult Congenital Disease Project Board's 'Service model for the delivery of care to Adults with Congenital Heart Disease in North West England, Isle of Man and North Wales' is currently being finalised. For more details please contact:

Either
Dr Su Sethi E-mail: Su.Sethi@clha.nhs.uk

or
David Stockdale E-mail: David.Stockdale@clha.nhs.uk
44. The following population tables have been produced by Department of Health analysts as a guide for commissioners using a formula based on prevalence and survival rates.

Adults with congenital heart disease: prevalence by age by Strategic Health Authority by severity

45. The assessment of prevalence starts out with data on the birth prevalence and survival rate to the year 2000. The data is taken from tables 1-3 of the report of the 32nd Bethesda Conference – Care of the Adult with Congenital Heart Disease.¹
46. The Bethesda Report tables provide information on birth prevalence by three categories of severity – complex, moderate and simple – and survival rate to 2000 by year of birth.

1 Webb GD, Williams RG. 32nd Bethesda Conference: "Care of the Adult with Congenital Heart Disease". J Am Coll Cardiol 2001;37:1162-1198.

- 47. The straightforward method of estimating prevalence for 2000 is to apply these survival rates to each year’s live births. There are two drawbacks. Data on live births are only readily available for England and Wales, not England. Secondly, the method takes no account of net migration.
- 48. However, assuming that England’s share of the England and Wales population is 94%, the estimates for 2000 using this straightforward method are as follows:

Adults with congenital heart disease: prevalence by severity

Complex	Moderate	Simple
16,878	49,254	63,257

- 49. As an **alternative** method of estimating, for example, the number of births in 1940 corresponding to the sixty year old population in 2000, the population can be grossed up by an estimate of the proportion of the 1940 birth cohort surviving to 2000. This rate is derived from a survival curve based on the relevant set of historical annual death rates. For example, for those born in 1940, their mortality rate at 20 would be the rate in 20 year olds in 1960, and so on.
- 50. This method presupposes that immigrants would have been subject to the same mortality rates at comparable ages as the population born in this country. This assumption may well not hold good, but there is no obvious way of taking this into account.
- 51. The estimate of prevalence in 2010 is complicated by the absence of forecasted survival rates. The approach taken here marks-up the most recent all-cause population death rates for each age by a series of ratios calculated using age band and severity congenital heart disease death rates from the Bethesda report and appropriate contemporary all-cause death rates. Ten-year survival rates (2001 to 2010) are calculated from the marked-up death rates for each age, and then applied to the 2000 adults with congenital heart disease prevalence estimates, again by age, to give 2010 prevalence.
- 52. The “mark-up” ratios gain their numerators from survival rates for each age band and severity. First year survival is excluded as being unrepresentative of subsequent years. Denominators are the all cause death rates from second year after birth to 2000 for the median age in each of the Bethesda age bands. This method is necessarily crude but at least provides rough points of reference regarding the likely scale of the increase in adults with congenital heart disease by 2010.

53. The estimated prevalences in 2000 and 2010 by SHA and severity, using this alternative method and taking account of any differences in age structure and gender ratio, are as follows:

	Complex		Moderate		Simple	
	Prevalence in 2000	Prevalence in 2010	Prevalence in 2000	Prevalence in 2010	Prevalence in 2000	Prevalence in 2010
Norfolk, Suffolk & Cambridgeshire	710	890	2190	2640	2840	3310
Bedfordshire & Hertfordshire	540	690	1650	2020	2140	2520
Essex	520	670	1610	1970	2100	2460
North West London	730	830	2040	2360	2640	2970
North Central London	490	570	1360	1590	1760	2000
North East London	590	730	1650	2010	2120	2490
South East London	580	690	1640	1950	2120	2450
South West London	510	590	1440	1690	1870	2130
Northumberland, Tyne & Wear	460	570	1410	1690	1820	2110
County Durham & Tees Valley	380	480	1160	1410	1490	1750
North & East Yorkshire & Northern Lincolnshire	520	660	1620	1970	2110	2460
West Yorkshire	730	920	2140	2630	2770	3260
Cumbria & Lancashire	610	790	1890	2320	2450	2880
Greater Manchester	880	1110	2600	3180	3360	3950
Cheshire & Merseyside	770	980	2360	2880	3050	3580
Thames Valley	740	920	2220	2680	2880	3350
Hampshire & Isle of Wight	600	750	1820	2200	2360	2740
Kent & Medway	510	660	1580	1950	2050	2420
Surrey, Sussex	800	1010	2510	3030	3260	3800
Avon, Gloucestershire & Wiltshire	730	910	2210	2670	2870	3340
South West Peninsula	480	610	1520	1840	1980	2310
Dorset & Somerset	360	460	1130	1380	1470	1720
South Yorkshire	430	540	1290	1560	1660	1950
Trent	870	1100	2670	3240	3470	4040
Leicestershire, Northamptonshire & Rutland	540	680	1630	1980	2100	2460
Shropshire & Staffordshire	480	610	1500	1820	1950	2270
Birmingham & the Black Country	780	1000	2280	2830	2930	3480
West Midlands South	500	640	1550	1880	2020	2350
Total	16870	21030	50680	61390	65640	76570
Growth to 2010		20%		17%		14%

Special needs population

54. A further consideration in planning services is the high number of learning disabled adults with a congenital heart condition in particular those with Down's Syndrome. Down's Heart Group (UK) estimates that:
- In every 700 babies born, there is likely to be one with Down's Syndrome.
 - About 40% of the babies born with Down's Syndrome will have a heart problem.
55. There are no official statistics on the number of people with learning disabilities in England. A recent study² suggests that 2% of the general population has a learning disability and a further study³ predicts that by 2021 there will be a 37% increase in the number of adults with learning disabilities.

Facilities and skills in specialist and local GUCH centres

56. The following list has been agreed by the External Reference Group and informed by documents cited in paragraph 41.

Specialist centres

Specialist centres for young people and adults provide care by a multi disciplinary and multi speciality team comprising:

Cardiologist, specialist in adult congenital heart disease
Cardiologist – interventionist with specialist training in congenital heart disease
Cardiologist – electrophysiologist with specialist training in congenital heart disease
Cardiologist – imaging (Echo and Magnetic Resonance Imaging)
Congenital cardio thoracic surgeon (preferably based on site)
Nurse specialising in service for adults with congenital heart disease
Anaesthetists who regularly undertake anaesthesia for people with congenital heart disease
Radiologist with understanding of congenital heart disease
Perfusionist
Dental surgeon
Obstetrician and fetal medicine specialist (as required)
Clinical psychologist
Physiotherapist
Dietician
Social worker
There is appropriate on call cover and expertise.

2 Estimating the current need /demand for support for people with learning disabilities in England Institute for Health Research, University of Lancaster, Emerson E, Hatton C,2004

3 Estimating the future needs /demand for support for people with learning disabilities in England Institute for Health Research, University of Lancaster, Emerson E, Hatton C,2004

The centre will have the following facilities:

- A ward where staff have special expertise in adult congenital heart disease
- An intensive therapy unit with relevant expertise
- A specialist outpatient service
- Emergency care
- Specialist obstetric care including fetal medicine

It will be able to access the following diagnostic/treatment services:

- Blood testing
- Electrocardiogram (ECG)
- Chest X ray
- Exercise testing
- Echo including fetal echo and transoesophageal echocardiography (TOE)
- Magnetic Resonance Imaging
- CT scanning

It will have established links (i.e. referral criteria, patient pathway and clinical management protocols) to other services, which may not necessarily be on the same site as follows:

- Transition service with a paediatric cardiac centre
- Local GUCH centres
- Specialist dentistry
- Heart transplantation services
- National Pulmonary Hypertension Service
- A genetics service
- Communications with staff in services for people with learning disabilities
- Special needs advice for anaesthesia

It will undertake data collection to enable clinical audit of all patients within the catchment area.

Local GUCH centres

57. A local GUCH centre may be part of an adult tertiary cardiac centre or a cardiac department in a DGH. They provide the following:
- A cardiologist with a special interest in adult congenital heart disease (see paragraph 64)
 - Emergency treatment of heart failure and arrhythmias
 - Joint clinics with visiting specialist cardiologists
 - Immediate access to a specialist centre for advice

It will be able to access the following diagnostic/treatment services:

- Blood testing
- ECG
- Chest X ray
- Exercise testing
- Echo

Local GUCH centres will have access to psychological and social worker support.

58. After consultation with a consultant specialist in adult congenital heart disease, the local GUCH centre may also provide:
- Non-cardiac care (medical and surgical)
 - Care associated with pregnancy, dental treatment or care requiring a general anaesthetic
 - Assessment of risk for non-cardiac conditions
59. The GUCH Patients' Association carries out a survey of specialist centres based on these definitions and the indicators of high quality care. Results summarise information provided by the centres. The first set of results will be available on the GUCH Patients Association website in early Summer 2006 at the following address: <http://www.guch.org.uk/>

Transition services

60. The aims of a transition service are to ensure uninterrupted and coordinated care, to promote understanding of the patient's condition and its impact on adult life (higher education, employment, relationships), to develop skills in self-care and promote independence and to support parents. An example of the key elements of a transition service has been provided by Birmingham Children's Hospital and University of Birmingham Hospitals Trust.

- 'Transition for Young People at Birmingham Children's Hospital: The Cardiac Unit'.

For more details contact:

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Other guides are available to support young people and their parents:

- 'Information for GUCH Teenagers' – GUCH Patients' Association website
- British Heart Foundation/GUCH Patients' Association booklet 'Living with congenital Heart Disease' Information and support for teenagers and adults

Also from Birmingham Children's Hospital and University of Birmingham Hospitals Trust:

- 'Grown Up Congenital Heart Clinic (GUCH), a Guide for Young People'
- 'Growing up with a Congenital Heart Defect or Heart Disease' – a guide for parents
- 'Moving on to adult hospital care' – a guide for young people

Workforce and training

61. The British Cardiac Society Working Party report on the cardiac workforce provides the profession's view on requirements and has useful modelling to inform the development of local workforce plans.

62. The Congenital Heart Surgery Committee of the European Association of Cardio-Thoracic Surgery (EACTS) and European Heart Surgeons Foundation make recommendations on volume/ staffing levels. These are available at: <http://ejcts.ctsnetjournals.org/cgi/content/full/24/3/343>

63. The Joint Committee on Higher Medical Training has published its latest curriculum for cardiology in August 2005. This sets out what is expected of **all** cardiology trainees in terms of objectives, knowledge, skills and attitude for adults with congenital heart disease.

64. In the advanced curriculum for adults with congenital heart disease the competencies of the two types of cardiologist are set out in more detail. Type 1 are those who practice as a specialist in a specialist unit which includes specialist surgeons. Type 2 are those who practice cardiology with a special interest in adults with congenital heart disease in non-specialist units (local GUCH units). Other sub specialists could undertake the type 2 training module to help their knowledge and skills base if appropriate.
65. In accordance with 'Good Medical Practice' guidance published by the General Medical Council, doctors are expected to:
- recognise and work within the limits of their professional competence;
 - be willing to consult colleagues;
 - be competent when making diagnoses and when giving or arranging treatment.
66. The curriculum also sets out an expectation of what an advanced training centre for adults with congenital heart disease should have.

It is available on the Joint Committee on Higher Medical Training website <http://www.jchmt.org.uk/cardio/index.asp>

List of skills and knowledge areas for nurse specialists

67. As services expand to meet the needs of a growing adult population, the demand for training and development of nurses will increase for both nurses on cardiac wards and nurse specialists in congenital heart disease. To assist in this development, the External Reference Group in consultation with nurse consultants has drafted a minimum set of skills and knowledge areas for ward based nurses, senior ward managers and nurse specialists. Exploratory discussions have taken place with a view to using these skills and knowledge areas to develop competencies for nurse specialists as part of the 'Skills for Health' programme. These will need to link to the NHS Knowledge and Skills Framework.

Adult Congenital Heart Disease post registered nurse – suggested areas of knowledge

68. These suggested areas of knowledge for nurses caring for adult patients with congenital heart disease are intended to enable discussion regarding potential demands for training and development as services expand to meet the needs of an increasing population. It will be important to support learning through placement experience.
69. Clearly, parallel to the development of any skills or knowledge competencies would be the development of an agreed academic framework, reflecting the proposed changes to the Nursing and Midwifery Council levels of registration.

Three types are suggested:

Cardiac ward based staff

Senior cardiac ward manager

Adult Congenital Heart Disease/GUCH nurse specialist

The objective is to be able to conduct physical and psychological nursing assessment and care of young people and adult patients with congenital heart disease.

Cardiac Ward based staff knowledge/skills/attitude

- Normal anatomy and physiology of the cardiovascular system
- Basic embryology and morphology
- Understand altered anatomy of the heart and great vessels and effect
- Understand effects of altered anatomy on cardiovascular system (CVS) observations
- Knowledge of common congenital heart defects
- Knowledge of interventions, procedures and treatment of common congenital heart defects
- Basic assessment of the cardiovascular system
- Knowledge of effects of cardiopulmonary bypass
- Understand the potential hazards involved in use of oxygen therapy
- Understand monitoring and basic ECG interpretation
- Ability to teach patients and their family about their condition, investigations and treatment
- Ability to teach patients and family about lifestyle related issues
- Ability to teach colleagues
- Be able to care for patients with learning difficulties who have congenital heart defects and support their families
- Understand the psychological effects of congenital heart disease
- Understand the effects of the transition process
- Communication skills including communicating with young people
- Knowledge of relevant pharmacology

Experienced cardiac ward manager knowledge/skills/attitude

Including all of the above plus:

- Understanding of a broader knowledge of complex congenital heart defects
- Understanding of abnormal anatomy/physiology of the heart
- Understanding of the implications of pregnancy in women with congenital heart disease
- Understanding contraceptive therapy for women with a congenital heart disease
- Understanding of diagnostic tests
- Understanding of specific complications or problems which this group of patients might develop
- Understanding of Arrhythmias/Sudden Cardiac Death and Chapter 8 of National Service Framework chapter
- Understanding of bridge to transplant/transplantation
- Understanding of the numbers of adult patients with congenital heart disease in their Trust and the potential demands on the service
- Ability to teach staff at all levels in the clinical area
- Knowledge of support agencies for patients with specific conditions, syndromes or learning difficulties
- Knowledge of specific health promotion issues e.g. family planning, diet/weight school/work life/exercise/insurance issues/benefits/driving/travelling
- Understanding of palliative care support

Adult Congenital Heart Disease/GUCH nurse specialist knowledge/skills/attitude

- Lead the development of clinical nursing practice for GUCH patients.
- All of the above (previous two categories) plus:
- Plan own caseload management within clinical areas and clinics
- Develop in-house teaching programs for staff caring for GUCH patients
- Establish links with local higher education establishments and academic colleagues to explore formal education provision opportunities for staff wishing to develop their knowledge and expertise further

- Develop skills in utilizing and undertaking research to enhance evidence-based practice. This could include publishing and presenting at conferences
- Provide clinical expertise to relevant managers to ensure an efficient, effective, quality service
- Explore the need for ‘hub-and-spoke’ links with nurses within secondary and primary referring centres to enhance the patient journey
- Provide individualised information and support to patients and their families
- Provide information and support to staff in clinical areas caring for adults with congenital heart disease and their families
- Counselling skills
- Awareness of own support needs

70. The exact nature of each specialist nurse post will depend on the requirements of individual centres. However, it is envisaged that there will be an increasing demand for individuals to undertake roles previously undertaken by medical colleagues, such as:

- Interpretation and ordering of non-invasive investigations.
- Advanced health assessments
- Implementing nurse led clinics
- Developing enhanced counselling skills
- Develop links with psychology services
- Receive and make referrals from and to appropriate support services (e.g. social services, dietician, physiotherapy etc.)

Annex A: External Reference Group Membership

Dr John Gibbs	Consultant Paediatric Cardiologist, Leeds General Infirmary
Dr Simon Gibbs	Consultant Cardiologist, Imperial College School of Medicine, Hammersmith Hospital
Dr Sara Thorne	Consultant Cardiologist, University Hospital Birmingham NHS Foundation Trust
Michael Cumper	Chairman of Trustees GUCH Patients' Association
Sheena Vernon	Adult Congenital Heart Nurse Specialist, United Bristol Healthcare NHS Trust
Prof. John Deanfield	Professor of Cardiology, Great Ormond St Hospital
Mr Victor Tsang	Consultant Cardiothoracic Surgeon, Great Ormond St Hospital
Mr Marcus Haw	Consultant Cardiothoracic Surgeon, Southampton University Hospitals
Cliff Lake	Chair, Down's Heart Group
Dr Roger Davies	Consultant in Special Needs Dentistry Eastman Dental Hospital and Institute

Annex B: Glossary

Anticoagulation – thinning of the blood with drugs, usually warfarin.

Congenital heart disease – heart disease caused by deformed development of the heart in the womb.

Echocardiography – use of ultrasound to provide moving images of the heart muscle and valves and make measurements of the functions and any damaged tissues.

Electrocardiogram (ECG) – electrical tracing of the heart's activation recorded on the body's surface.

Healthcare Resource Groups (HRGs) – standard groupings of clinically similar treatments, which use common levels of healthcare resource. They may be considered as 'units of currency' within the health service, allowing for costings across services.

International normalized ratio (INR) – a method of standardising prothrombin time results (see below), developed to compare prothrombin time results between laboratories using different test methods.

Magnetic Resonance Imaging (MRI) – a noninvasive diagnostic technique that produces computerized images of internal body tissues and is based on nuclear magnetic resonance of atoms within the body induced by the application of radio waves.

Neonates – newborn infants.

Prophylaxis – preventive treatment.

Prothrombin time (PT) – a blood test that measures how long it takes blood to clot. PT is used to monitor treatment with medication that prevents the formation of blood clots.

Tertiary cardiac centre – centre providing specialised cardiac services including surgery. Not all tertiary cardiac centres provide specialised management of adult patients with congenital heart disease, paediatric cardiology or complex electrophysiology.



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