

Adult Congenital Heart Disease (ACHD) in Yorkshire and the Humber: A briefing document

Introduction

The vast majority of children operated upon with congenital heart problems now survive into adult life. As such there is now a large (and growing) population of young (and older) adults with congenital heart disease across the country who require ongoing specialist care for their heart problem.

Current estimates indicate there are more than 13 000 patients over the age of 16 years with congenital heart disease now living within the Yorkshire and Humber area. This figure is expected to increase overall by around 20% between 2007 and 2015. The number of patients with very complicated congenital heart problems is expected to increase by 80% in the same time period. Overall patient numbers are expected to continue to rise year on year until 2040. To put these numbers into context, the Department of Health's own estimates (GUCH guide) indicate that currently in the UK, the numbers of adults with congenital heart disease **outnumber** the number of children.

The huge growth of the adult congenital heart disease population has meant that the tertiary services for these patients in the region, run from the Leeds General infirmary have developed significantly over the past decade. There has been considerable investment in cardiologists with special training in adult congenital heart disease (currently 3 in Leeds) and all of the services needed to support them. These developments have been driven by an over-riding philosophy that these patients are best cared for by multi-disciplinary teams within units that offer as much care as they possibly can for these patients under one roof. In Leeds there is seamless care for patients with congenital heart disease from pre-natal diagnosis of the problem to care during childhood and transition into adult life. Whilst understanding and respecting that these patients are different from children, we also feel that there are elements of their care better delivered by adult congenital teams in close proximity with paediatric cardiology and cardiac surgery colleagues who may have treated patients for many years. In addition under one roof adults with congenital heart disease can access almost every other services they require e.g obstetrics, fertility, renal, respiratory medicine, acquired cardiology and cardiac surgery etc, etc, etc.

Key factors for running a successful ACHD service

There are already national guidelines for running a safe and successful ACHD service outlined in the 2006 DH document entitled "A commissioning guide for services for young people and Grown Ups with Congenital Heart Disease (GUCH)" (known as the "GUCH guide"). This document was a consensus of the opinions of UK professionals working in the area along with patient groups and commissioners. In the Yorkshire region we have taken a very proactive approach in implementing the recommendations of this document. In summary, with recommendations from the "GUCH guide" highlighted:

- 1) **"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"**.

“A nurse specialist is available as the main contact point for each patient in transition. “

Current situation in Leeds: There is a transition system in Leeds with a specialist nurse and cardiologist. Dedicated teenage in-patient accommodation is available within the paediatric cardiac unit. Physically these patients can be cared for within the same hospital regardless of age and so there is not the same requirement for separate transition wards etc as there would be within a stand alone childrens hospital.

***NOTE:** Whilst there is general recognition that complex patients with congenital heart disease are best cared for in units providing all services under one roof as in Leeds regardless of age but with age appropriate care, the majority of regions in the UK do not have hospitals of this nature available. Thus the recommendations of the GUCH guide apply differently depending on the local resources.*

- 2) **“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.”**

“Specialist centres have shared care protocols with local GUCH centres and GPs. This may include outreach services.”

“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.”

“Accommodation for families /carers is available for patients who need an inpatient stay.”

Current situation in Leeds: All ACHD patients are given hand held patient records with details of their condition and local/national information sheets. The patient held record contains contact information and all patients are copied correspondence relating to their case which are stored in the patient held record for reference. Local GP’s/cardiologist/local GUCH centres are routinely copied into correspondence. For a number of “high risk” cases we provide additional information including a copy of the patients electrocardiogram (ECG) in case of an arrhythmia which can be shown to the local ambulance/A+E and an “open access” letter, agreed with the Yorkshire ambulance service allowing certain patients to be brought directly to Leeds in the event of an emergency bypassing local hospitals if deemed necessary. Patient information is delivered in a manner in which patients can understand it regardless of disability.

- 3) **“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.”**

Current situation in Leeds: See 2 above. All cases are offered an appointment with a GUCH specialist cardiologist (3 specialist cardiologists in Leeds as defined by the British congenital cardiac association document). Sometimes the appointment is in Leeds or sometimes in our outreach clinics depending on patient preference. In all cases a GUCH specialist is present.

4) **“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.”

Current situation in Leeds: We run an outreach service in Sheffield with a local GUCH cardiologist and plans are advanced for a second outreach clinic in Hull to start in the near future. We have very close links with paediatric cardiac services being within the same block of the hospital in the LGI. The GUCH team will see the paediatric cardiac team on a daily basis. We have formal links for shared care of appropriate patients with pulmonary hypertension (Sheffield) and transplnatation (Newcastle). Genetics services are available on site.

Congenital cardiologists are available 24/7 for advice to local teams.

5) **“Specialist centres provide teaching and training including training for technicians and also post-graduate education.”**

“Specialist centres provide a 24-hour point of reference for colleagues in ocal GUCH centres, tertiary centres, DGHs and primary care including general dental practitioners and community dental clinics.”

“Specialist centres work collaboratively with each other and local GUCH centres to ensure national audit. “

“Specialist centres provide teaching and training including training for technicians and also post-graduate education.”

Current situation in Leeds: We host “network meetings” for staff of all types on a 2 yearly basis during which there is a clinical education and also time to discuss network management and commissioning requirements. In addition we have a fully functioning website (unique as far as we are aware in the UK in terms of its breadth and content: www.yorksandhumberhearts.nhs.uk/default.aspx?id=271) containing a huge amount of information for patients, carers and professionals.

We run the only UK “core curriculum course” for adult acquired cardiology trainees to ensure that they get proper basic training in ACHD.

Congenital cardiologists are available 24/7 to local clinical staff for advice.

We submit data for national audit and encourage local centres to do this also.

6) **“Patients receive ongoing support and advice from a specialist GUCH nurse.”**

Current situation in Leeds: We have 2 GUCH specialist nurses and a plan for a 3rd.

- 7) **“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.”**

“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.”

Current situation in Leeds: We are attached to a dental school and have dentists available for help and advice. Advice is given to patients with regard to seeking dental advice but since the GUCH guide was written there have been changes to the provision of UK dentistry that have affected the access of patients to dentists.

- 8) **“Pregnant patients with complex cardiac conditions are managed at a specialist 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. “

Current situation in Leeds: A high-risk cardiac pregnancy clinic has been in place in Leeds for 65 years fulfilling all of these requirements.

Uniquely there is on site fetal medicine and reproductive medicine in the LGI.

- 9) **“Accommodation for families /carers is available for patients who need an inpatient stay.”**

Current situation in Leeds: Available to families/carers

ACHD services and the current review of paediatric cardiac services: the view of local clinicians

In 2008 the Department of health embarked upon a process aimed at reducing the number of centres in the UK performing children's heart surgery from 11 down to a smaller number (the "safe and sustainable" review as it has become known). The drivers for these proposed changes are fully outlined on their website:

www.specialisedservices.nhs.uk/safe_sustainable/childrens-congenital-cardiac-services

In essence the main reasons for the proposals were a perception that centres doing larger numbers of operations have better outcomes than smaller centres and that the small number of paediatric cardiac surgeons in the UK need to be working in larger groupings to ensure adequate rest and better results.

As is often the case with reviews of this nature there was an initial under-estimation of the "ripple" effect of moving a highly specialised service. During the conduct of the review it has become clear that moving paediatric cardiac surgical services will affect almost every element of hospital based paediatric care in the UK to some degree. Some of the effects will be relatively minor but in many cases, e.g. the provision of children's intensive care, there are likely to be major effects on the ability to provide other elements of a "sustainable" service in some areas of the country.

The enlarging adult congenital heart disease population in the UK also relies on the skills of paediatric heart surgeons to perform cardiac operations that are not infrequently required in adult life. In addition, although many adult patients can be treated by cardiologists with keyhole procedures thereby avoiding open heart surgery, these procedures can only be carried out with the support and cover of a paediatric heart surgeon within the same hospital. It is clear therefore that care of patients with ACHD beyond routine outpatient work can only take place in centres retaining the skills of paediatric cardiac surgeons. In effect therefore the paediatric cardiac surgical review has become a review not only of paediatric cardiac surgery or even paediatric cardiac care but a review of "specialist" ACHD services as well.

The British congenital cardiac association, the key body representing professionals caring for patients with congenital heart disease recognised the implications of the review of paediatric cardiac services for ACHD services and lobbied the department of health decision makers early in the review to consider ACHD within the remit of the "safe and sustainable" review given that the implications were so far reaching. This was refused, the review directors feeling that ACHD was such a big issue that it could not be reasonably dealt with in this workstream. Virtually every clinician involved with the care of these patients was unhappy with this decision and this was made very clear to the review team at the DH.

For reasons that remain unclear to clinicians at the very last minute the review team suddenly decided to conduct a review of ACHD services outside and in parallel to the paediatric cardiac surgical review. The first meeting of this review team was 2 months ago. As a result we are now in a position where the paediatric cardiac service review which has been ongoing for over 3 years plans to report with its recommendations within the next 3 months, just as an ACHD review is in its infancy!

Whilst on the surface of it one might feel encouraged that the DH team have at last "listened" to clinicians with regard to ACHD there are serious and very obvious problems. It is quite clear from the description above that an ACHD specialist unit (like

the one in Leeds) cannot exist without paediatric cardiac surgery on site. Therefore if this service is removed in effect the ACHD service in Leeds automatically will be “downgraded” to a local centre. This will result in the loss of many if not all of the skills available locally and the ACHD patients of this region will face the same prospect that is facing families with children with congenital heart disease i.e. long travel times to other areas of the country for care.

Ultimately if the “Safe and sustainable” review had considered ALL aspects of the care of patients with congenital heart disease equally then the process would have at least been fair. It is clear however that those patients who are unfortunate to be above 16 years of age have been effectively disenfranchised from the “real” decision making review whilst the views and needs of younger patients (and their families) affected with the same disease process have been encouraged to help shape the review. This is clearly totally inequitable and unfair. The key issue is that the new review of ACHD services can only reach one conclusion once the paediatric cardiac surgical review has given its decision as ACHD specialist centres will have to go where the childrens heart surgeons are placed. Both reviews are therefore flawed and in our view, no matter how painful it may seem the process should be repeated and performed properly and fairly if it really to succeed in its aims of providing care to all of these patients that is really “safe and sustainable”.