

Original Article

Adult congenital heart disease interventions: recommendations from a Joint Working Group of the British Congenital Cardiac Association, British Cardiovascular Intervention Society, and the British Cardiovascular Society

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Abstract In order to optimise care of the adult patients with complex congenital heart disease, there is a need to develop recommendations for interventions. This document is the work of representatives of the three relevant societies and provides recommendations for institutions and operators performing cardiac interventions in these patients.

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THIS DOCUMENT PROVIDES RECOMMENDATIONS FOR institutions and operators performing cardiac interventions in Adult Congenital Heart Disease in the United Kingdom. These are based both on published evidence and on a consensus of experienced practitioners in adult and paediatric interventional cardiology and also take into account recommendations from previous reports.^{1–5}

This document is aimed at management of adult patients with congenital heart defects, defined as patients 16 years or older. The objective is to optimise patient care by recommending the requirements for services providing interventional management.

There is considerable variation in the level of complexity of lesions in different patients with Adult Congenital Heart Disease. The skills needed to look after infants or children with complex lesions are different from those needed to treat an adult patient with a patent foramen ovale. Children with complex lesions who have undergone complex treatments need to be cared for in adulthood by those with the necessary skills and expertise. Skills from the paediatric and adult cardiology communities may need to be combined for the optimal care of some complex cases. Whereas there is a growing number of paediatric cardiologists who have now taken on the role of Adult Congenital Heart Disease specialists, it is also recognised that many adult cardiologists have undergone the appropriate training to identify and treat a range of less complex lesions (outlined below).

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The British Congenital Cardiac Association, British Cardiovascular Intervention Society, and British Cardiovascular Society need to (a) work with the Department of Health and commissioners to develop a consensus view on how such interventional treatments should be delivered on a national scale, (b) encourage and help organise appropriate training in the relevant procedures, (c) develop the standards needed to be recognised as an Adult Congenital Heart Disease interventionist, and (d) encourage units to collect validated audit data and then oversee an informed interpretation of analyses of such data. The means by which these four aims are to be achieved require further discussion, but this document should encourage clinical networks and commissioners to initiate regional discussions. The three professional societies are committed to working with National Institute for Health and Clinical Excellence on appropriate treatment guidelines and technology appraisals. This document has been developed in consultation with the Grown-Up Congenital Heart Patients Association.

For any operative skill in medicine, there are three requisites:

- (i) a full understanding of the anatomical defects concerned, the related pathophysiology, and the means by which to make accurate diagnoses;
- (ii) the acquisition of the appropriate knowledge and skills needed to provide effective treatment (including an understanding of the complications of treatment and their management), taking into account comorbidities and complexity of disease;
- (iii) a referral practice that provides sufficient cases to ensure that skills are maintained and developed.

There is insufficient clinical research to provide definitive guidance on determining a precise level of continuing activity to maintain skills. Nevertheless, there is agreement that regular practice is likely to produce better results than occasional practice. In this field, there are some relatively common procedures but other procedures occur infrequently. There are two approaches to the management of rare cases:

- (i) ensure that the patient is treated in a centre that has a relatively high volume of all aspects of Adult Congenital Heart Disease activity, such that those undertaking rare cases are most likely to have the appropriate skills to manage them and
- (ii) cross-referral between specialised Adult Congenital Heart Disease units such that rare cases are treated in higher volume by a small number of units.

This is a challenge not only to those adult interventional centres that carry out some of the more common and less complex procedures, but also to all recognised Adult Congenital Heart Disease centres.

This document does not deal with pacing/electrophysiology interventions in Adult Congenital Heart Disease patients. It is recognised that such procedures may require collaboration between electrophysiologists and interventional cardiologists with expertise in Adult Congenital Heart Disease.

Patients

Adult Congenital Heart Disease patients requiring intervention fall broadly into two categories:

Less complex disease

Typical lesions include patent foramen ovale and ostium secundum atrial septal defect. Usually, these lesions are isolated but it is important to understand that even isolated cardiac lesions have their anatomical complexities – single versus multiple or discrete versus fenestrated atrial septal defects – or may be associated with anomalies in other systems. Where therapeutic strategies include the use of transcatheter interventions, it is essential that teams establish the full extent of the abnormality and understand the anatomical variations they may encounter.

More complex disease

Such disease includes the large array of complex structural anomalies, either de novo or as modified by surgical or transcatheter interventions. Paediatric cardiologists will already have treated many of these patients, but new cases can present in adulthood. Previously, such patients were referred from paediatric units to specific adult cardiologists, who had developed an interest in congenital heart disease. It is now recommended that clinicians with interests in paediatric and adult congenital disease should look after these complex adult patients in specialist Adult Congenital Heart Disease (or “Grown-up Congenital Heart”, or “GUCH”) clinics. However, such Grown-up Congenital Heart clinics are currently not available in all hospitals and often patients have shared care between local and specialist Adult Congenital Heart Disease services.

Patients with more complex Adult Congenital Heart Disease may develop acquired heart disease with increasing age. Adult cardiologists will therefore be involved in the management of many of these patients, and should be able to recognise congenital abnormalities and the issues raised by them. Where necessary, the patients should then be referred to a Specialist Grown-up Congenital Heart Centre.

Need for collaboration between a range of services

The population of patients with Adult Congenital Heart Disease is continuing to increase because of

improvements in the treatment of congenital heart disease in infancy and better treatment as adults in centres specialising in their treatment. Indeed, there are now more adults than children with congenital heart disease.⁶ Although survival has improved, many of these patients may develop complications related to the underlying heart defect or to previous surgery or transcatheter intervention, and may need further treatment.

Most new patients are initially referred to their local general hospital for initial assessment and diagnosis before being referred to specialised services. Many patients already known to the Grown-up Congenital Heart services may present to local hospitals as medical emergencies and thus these patients should be known to their local hospital.^{7,8} Expertise in dealing with Adult Congenital Heart Disease patients at the local hospital is likely to be less than at a specialist centre, and all cardiologists dealing with an unselected cardiology workload must be aware of the problems that beset these patients and be able to contact a specialist centre at all times. Many acute problems – such as treatment of acute heart failure, arrhythmias, etc. – can be dealt with immediately in the local setting, but further investigation to understand why a patient has presented will need additional expertise.

Indications for intervention

The development of interventional techniques, whether used in an elective or emergency setting, may replace the need for cardiac surgery in many cases. In general, the indications for intervention in congenital heart defects are similar to the surgical indications, although the threshold for intervention may be lower given the potential for reduced procedural risk. However, longer-term outcomes remain uncertain. Such uncertainties should be explicit in discussions with patients. Where there is a range of therapeutic options, or uncertainty about whether an intervention is justified, the decision for the intervention should be taken after discussions within an appropriate multidisciplinary team (see Sections “Requirements for all centers” and “Additional requirements for Specialist Adult Congenital Heart Disease centers”).

Interventional centres

This document considers two distinct types of services for Adult Congenital Heart Disease. A Specialist Adult Congenital Heart Disease service provides a comprehensive range of services for patients with Adult Congenital Heart Disease, including on-site specialist congenital heart disease interventions and

surgery. Local Adult Congenital Heart Disease services contribute to the care of patients with Adult Congenital Heart Disease, but offer a limited range of services including interventions and do not have on-site congenital heart disease surgery.

In contemporary practice, most interventional cardiology services treating adult patients concentrate expertise for non-congenital structural heart lesions in the hands of a relatively small number of adult interventional cardiologists. Their workload may incorporate established and new cardiac interventions – for example, transcatheter valve interventions, left atrial appendage occlusion, closure of para-prosthetic leaks, and embolisation of fistulas, but many also treat patients with the less complex Adult Congenital Heart Disease lesions. This document does not mandate that all interventionists with a structural heart disease interest also carry out Adult Congenital Heart Disease interventions, nor does it mandate that Adult Congenital Heart Disease interventionists carry out non-congenital structural heart procedures. Any interventionist – whether trained as a “paediatric”, “Adult Congenital Heart Disease”, or “adult” interventionist – performing Adult Congenital Heart Disease or structural heart interventions should, however, have the appropriate training and competencies to carry out procedures safely and effectively. Each service should decide which interventionists carry out specific procedures to ensure a sustainable programme. By necessity, this means that many coronary interventionists will not perform structural heart or congenital interventional work, and many paediatric or congenital heart interventionists will not undertake non-congenital structural heart interventions in adults. Given the relatively small number of cases that present for treatment on a yearly basis, it is also evident that many interventional services should not undertake structural heart or congenital interventional work.

There is consensus that skills required for any cardiac intervention are honed through practice and experience. The effect of the learning curve needs to be minimised by appropriate training and proctorship – see section “Training, supervision and proctoring” – both for trainee operators and for established operators learning new procedure skills. Established operators and institutions should carry out a sufficient procedural volume to maintain competence. If the volume is insufficient, operators and institutions should review the need for a local service, enter discussions with the regional or supra-regional specialised Adult Congenital Heart Disease centre and commissioners, and decide on appropriate changes to service configuration. This might involve merger of services or referral to colleagues with the expertise and institutional practice to maintain a high level of care for patients.

The three professional societies (British Congenital Cardiac Association, British Cardiovascular Intervention Society, and British Cardiovascular Society) recommend that provision of Adult Congenital Heart Disease services should be agreed among all stakeholders to ensure that an appropriate number of procedures are performed by services and by individual interventionists, with the objective of maintaining a high level of care. For most Adult Congenital Heart Disease interventions, robust evidence of a relationship between procedural volumes and outcomes is lacking. Although there is some evidence of a volume–outcome relationship for percutaneous closure of atrial septal defect,⁹ it is not sufficient to support recommendations for all procedures. Nevertheless, for many procedures, it is likely that low-volume activity will jeopardise procedural outcomes, and concentration of such procedures into a relatively small number of services with a limited number of operators is an appropriate strategy. The recommendations in this document are based on consensus and suggest levels of activity that are likely to provide good care for patients. This strategy will be reviewed as further evidence about volume–outcome relationships in Adult Congenital Heart Disease interventions becomes available.

The Department of Health guide to adult congenital cardiac services and the Paediatric and Congenital Cardiac Services Review, drawn up in collaboration with the British Congenital Cardiac Association, state that only appropriately qualified and experienced cardiologists should treat congenital cardiac anomalies.^{10,11} Adult patients with repaired or unrepaired complex congenital heart disease, who require cardiac catheterisation and interventions, should have the procedure performed by an interventional cardiologist with established expertise and training in congenital heart disease.

Requirements for all centres:

- Interventional services should be available continuously throughout the year with agreed arrangements to cover holidays and professional leave.
- A full range of diagnostic imaging facilities including contrast and transoesophageal echocardiography, CT, and MRI scanning should be available.
- Expertise in the use of intra-operative transoesophageal echocardiography and/or intra-cardiac echocardiography should be available for many procedures, and the use of these imaging modalities in addition to radiographic guidance is recommended.
- Owing to surgical options for these patients and the occasional need for urgent cardiac surgery during interventional procedures – for example,

for management of pericardial tamponade – there should be on-site cardiothoracic surgery with surgeons who have appropriate expertise to perform the operation on the congenital heart lesion that is being treated.

- The number of operators performing interventions will depend on the size of the centre, but it is recommended that a minimum of two operators should provide the service.
- Interventionists should be trained and be proficient in dealing with the specific lesions they treat, and should arrange for proctoring as necessary when learning a new skill. In contemporary cardiology practice, no interventionist can be competent in all cardiac interventions; interventionists should recognise their individual areas of competence and limitations and be ready to refer patients to colleagues when other specific skills are required.
- Interventionists should be capable of recognising and managing the potential complications of the interventions they carry out – for example, device embolisation.
- Regular multidisciplinary team meetings should occur, at which cases may be discussed by a team including an experienced echocardiographer, an Adult Congenital Heart Disease specialist clinician,¹² and an interventional cardiologist with appropriate training in Adult Congenital Heart Disease interventional procedures. If one individual assumes all of these roles, at least one other cardiologist with a special interest in Adult Congenital Heart Disease should be present. Increasingly, those with expertise in magnetic resonance imaging should contribute.
- Data on procedures undertaken should be submitted to the Central Cardiac Audit Database.
- All centres with a sustainable level of activity should have succession or re-referral plans where an individual operator is near to retirement or planning to cease Adult Congenital Heart Disease interventions for other reasons. This may require specific training of other experienced interventional colleagues or a new appointment.

Additional requirements for Specialist Adult Congenital Heart Disease centres

The specific recommendations in the Paediatric and Congenital Cardiac Services Review, relating to therapeutic cardiac catheterisation for centres with a fully comprehensive combined paediatric and adult Adult Congenital Heart Disease programme, state that all interventionists treating Adult Congenital Heart Disease patients should carry out at least 40 therapeutic catheterisations for congenital heart disease per year and that no centre should perform

such procedures unless there is a lead clinician specialising in therapeutic catheterisation, who carries out a minimum of 80 such procedures per year.¹¹ The document further notes that submission of data to the national audit programme should be deemed a prerequisite to practice in this area, and that all patients with congenital heart disease, irrespective of their age should be given the opportunity of a consultation with a cardiologist with the necessary expertise. These recommendations were strongly supported by the patient support group for Adult Congenital Heart Disease patients (Grown-Up Congenital Heart Patients Association).

- The centre should have a full range of services for Adult Congenital Heart Disease patients, as outlined in the published commissioning guide.
- The service should be led by a trained Adult Congenital Heart Disease consultant, with involvement and support from paediatric cardiologists, congenital cardiac surgeons, cardiac anaesthetists, and intensivists.
- Multidisciplinary team meetings involving at least two involved clinicians and a congenital cardiac surgeon should take place regularly. Less frequent but regular regional meetings for all operators from both Specialist and Local Adult Congenital Heart Disease services are recommended.
- There should be a lead interventionist for the unit, who undertakes at least 80 interventional cases per year.
- There should be at least two operators capable of performing the interventional procedures.
- Given the procedural volume requirements for the lead interventionist and the need for at least one other operator, and accepting that many procedures will be undertaken by joint operators, it is recommended that such a unit will need to perform a minimum of 100 procedures per annum. For procedures where joint operators are involved, there should be a designated “first operator” and “second operator”. Such joint working should be recognised in job plans.

Specific requirements for Local Adult Congenital Heart Disease centres

Institutional volume should be sufficient to allow all members of the team, and not just the operators, to maintain proficiency in the care of these patients. It is recognised that many of the skills required for Adult Congenital Heart Disease interventions are generic but it is not appropriate to carry out specific interventions infrequently; rare interventions should be concentrated in Specialist Adult Congenital Heart

Disease centres. Development of new local interventional services should only take place after discussion with both the commissioning bodies and the specialist Adult Congenital Heart Disease intervention programme in order to ensure safe development of the programme along with appropriate governance arrangements.

- All Local Adult Congenital Heart Disease centres should have experience in structural heart interventions.
- The most commonly performed procedures are patent foramen ovale and atrial septal defect closure. Centres that plan to perform only these procedures should do a minimum of 30 procedures per year. Centres should not set out to perform patent foramen ovale closure alone. Any centre carrying out fewer than 30 procedures per year should discuss governance arrangements with their regional Specialist Adult Congenital Heart Disease centre and commissioners and consider referring cases to alternative higher volume providers.
- It is recognised that in an early phase of development, or during a period of regional reorganisation, fewer cases might be performed, but a programme should reach the minimum level of activity relevant to the case-mix proposed within 3 years of starting. In addition, the selection of where procedures are performed should be determined by discussions with all the relevant stakeholders. In some circumstances, it might be proposed that experienced operators work at more than one site – for example, a Specialist Adult Congenital Heart Disease centre and a nearby Local Adult Congenital Heart Disease centre – and a regional programme might allow a lower level of less complex Adult Congenital Heart Disease activity at one site – assuming appropriate surgical cover. The total activity for the programme should meet the minimum standards.
- Given the requirement for a minimum of two operators at any one centre, most centres will perform more than 50 Adult Congenital Heart Disease interventions in total, but where numbers are lower it is recommended that two operators perform procedures jointly. In these circumstances, it should be clear as to who the “first operator” and “second operator” is. Such joint working should be recognised in job plans.

Adult Congenital Heart Disease Interventionists

Interventionists should:

- have an understanding of the detailed anatomy of the congenital heart defects being treated.

- be able to demonstrate evidence of appropriate training in congenital cardiac disease¹³ and to demonstrate ongoing clinical professional development and continuing medical education – this could be in the form of attendance of at least one appropriate interventional meeting a year. The level of training and expertise should be sufficient to enable competence for the interventional procedures being undertaken.
- have knowledge and understanding of the indications and contraindications for interventions in Adult Congenital Heart Disease patients.
- have understanding of the potential complications of interventional procedures and their management, including use of retrieval devices.
- be familiar with the full range of equipment necessary for the interventional procedures they undertake in Adult Congenital Heart Disease patients.
- not undertake a procedure for which they are not specifically trained, unless proctored by an experienced operator, who should be approved by the company whose devices are to be used.
- work with colleagues and perform joint procedures wherever possible to ensure that the appropriate skills are developed and shared.
- attend multidisciplinary meetings and case conferences relevant to their case-mix.

Training, supervision, and proctoring

All operators should undergo a period of training and supervision by an experienced operator before carrying out an unsupervised Adult Congenital Heart Disease intervention. A learning curve is well recognised for the acquisition of any new skills, but can be minimised by appropriate training, which might include use of simulators, and proctorship. Setting clear numbers for proctored cases is problematic as different operators learn at different speeds and some operators may not be able to successfully develop the appropriate skills. It is appropriate for proctors to determine whether individual operators (with their supportive teams) have reached a level where they can operate independently. This may place responsibility on proctors to determine that a particular team has reached the necessary level of proficiency. The number of cases that need to be completed during the period of supervision will vary from operator to operator and should be determined by the proctor. Competence in performance of procedures should be the benchmark, rather than achievement of a minimum number of procedures. In some circumstances, the proctor may consider that the interventionist or the team is not proficient or able to operate independently, in which case the proctor has a responsibility to communicate

any concerns to the lead clinician for the service. Even when an operator is judged to be competent, it may be appropriate to consult with the former proctor.

Consent

Informed consent for any cardiac intervention is essential. Written information on the procedure should be provided to the patient, who should be given ample opportunity to obtain a further opinion in case of uncertainty. Whenever possible, the patient should be encouraged to involve those close to them in the decision-making process. Consent should also be obtained to allow for external validation of data submitted to the Central Cardiac Audit Database. The consenting process should otherwise conform to the Department of Health and General Medical Council recommendations.^{14,15} Patients should be informed when a procedure is being undertaken in the presence of a proctor.

Audit

The Care Quality Commission now includes the UK Central Cardiac Audit Database data in their assessments, when carrying out evaluation of services provided by a Trust.

- All Adult Congenital Heart Disease interventional services must commit to the requirements of the National audit process.
- Both Local and Specialised Centres should undertake regular audit of interventional procedures in accordance with the National Institute for Health and Clinical Excellence Interventional Procedures Guidance programme.
- Both Local and Specialised Adult Congenital Heart Disease services should be formally audited at least annually, and this should include review of interventional case volume, case-mix, and complications. Audit data should be shared with colleagues from referring centres.

Review of recommendations

Interventions in Adult Congenital Heart Disease patients are an evolving discipline and new data are emerging at a rapid pace. Therefore, these recommendations will be reviewed in 3 years.

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