

Recommendations for organization of care for adults with congenital heart disease and for training in the subspecialty of ‘Grown-up Congenital Heart Disease’ in Europe: a position paper of the Working Group on Grown-up Congenital Heart Disease of the European Society of Cardiology

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Received 23 January 2013; revised 11 December 2013; accepted 22 December 2013; online publish-ahead-of-print 27 January 2014

Introduction

The European Society of Cardiology (ESC) has recently published practice guidelines for the management of grown-up congenital heart disease (GUCH).¹ For the sake of space, this document did not include recommendations for the organization of care for adults with congenital heart disease (CHD) nor for training in the subspecialty of adult CHD, although emphasis was given to their importance and the need for a separate recommendations paper in the future.¹ This publication is a position paper from the ESC Working Group on Grown-up Congenital Heart Disease, which makes recommendations for the standards and organization of care for adults with CHD, as well as for training in GUCH within Europe. Obviously, there is a lack of solid scientific data to support such recommendations and the majority of them must remain based on expert consensus.

The remarkable improvement in survival of surgically and interventionally treated patients with CHD has led to an increasing number of

GUCH patients, in particular those with more complex disease. There are also patients with newly diagnosed CHD lesions in adulthood such as atrial septal defect, coarctation of the aorta, Ebstein's anomaly and congenitally corrected transposition of the great arteries. The 32nd Bethesda Conference report in 2001² estimated that there were ~2800 adults with CHD/million population, with more than half having moderate or highly complex disease. More recently, Marelli *et al.*³ reported a prevalence of 4090/million adults in Quebec for the year 2000 with 380 having severe disease. In Europe, we are currently faced with an estimated GUCH patient population of ~2.3 million, which significantly outnumbers the paediatric CHD population of ~1.9 million patients.⁴ There is general agreement that GUCH patients have very special needs and therefore physicians responsible of their care need specific expertise and training.^{1–11} It is particularly the growing number of adults with moderate and highly complex CHD who need such expert input. Although the majority of GUCH patients will have had surgical or catheter interventions, most cannot be considered cured but rather are palliated and

The opinions expressed in this article are not necessarily those of the Editors of the *European Heart Journal* or of the European Society of Cardiology.

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therefore have to live with a chronic disease. They require long-term surveillance and often re-interventions over their life-time for residual anatomical and/or functional abnormalities. The wide variety of congenital defects and their specific long-term problems as well as changing treatment strategies including medical, device, surgical, and interventional techniques remains a particular challenge when planning GUCH care. The organization of care and training programmes for those involved in care provision therefore needs to be defined, in order to meet the needs of this special population.^{1–11}

Organization of care

The 2003 ESC guidelines⁶ stratified patient care into the three levels by CHD diagnosis: (i) patients who require care exclusively follow-up in a specialist centre (level 3); (ii) patients in whom shared care can be established with the appropriate general adult cardiac services (level 2); and (iii) patients who can be managed in 'non-specialist' clinics (with access to specialist care if required; level 1). The current ESC guidelines¹ did not attempt to assign a hierarchy of care based by the underlying CHD lesion. Although complex defects can easily be assigned to level 2 and 3, even simple defects may require specialist care under certain circumstances (e.g. atrial septal defect with pulmonary arterial hypertension). It was therefore felt that the best model of care is one in which GUCH patients are seen at least once in a specialist centre and the GUCH specialist will then make recommendations for the level of care and follow-up intervals on an individual patient basis. In any case, a functioning network between specialist centres, local general adult cardiology departments, cardiologists in private practice (where available) and family practitioners is of critical importance.

Transfer from paediatric cardiology to GUCH care should occur by the age of 18 years, although an earlier transition process may occur in some countries depending on the local healthcare policies. A detailed medical report should summarize the course of disease including most recent as well as basic findings, all interventions and complications, and current clinical status. Particularly in cases of high complexity, it may be desirable that at the first visit to the GUCH centre or at last visit to the paediatric clinic, both the paediatric cardiologist and the GUCH cardiologist are present.

This document will define the requirements of a specialist GUCH centre. These supra-regional centres should serve as centres of

excellence that will provide care of the highest level. They will also provide education and training for subspecialization in GUCH, while promoting research and innovation. Such centres should serve a population of ~5–10 million people. Cardiologists and primary care physicians who take care of GUCH patients should establish a referral relationship with a specialist centre. This should include the provision of timely telephone advice, informal consults, rapid consultant review, as well as shared care arrangements for patient follow-up.

The definition of a specialist GUCH centre has previously been proposed by several national and international groups.^{2,4–10} The present recommendations try to incorporate these proposals as well as current working models of GUCH care within Europe. It must, however, be stated that these recommendations are not evidence based due to the lack of scientific data and are therefore based on expert consensus.

Institutional requirements

Specialist GUCH centres should ideally be co-located within a setting where there is adult and paediatric cardiology and congenital heart surgery. If GUCH services and paediatric services can for country specific reasons not be co-located they must at least have a strong joint transition programme and joint multi-disciplinary teams with defined care pathways and proven collaborations both clinically and academically. The entire spectrum of adult medicine should also be available (see Supplementary material online, *Table S1*). It is essential that there are strong links and defined referral pathways with the department of obstetrics and gynaecology for pregnancy care in women with congenital heart disease, but as there is also an increasing frequency of cardiac and non-cardiac co-morbidities in the ageing GUCH patient population, there must be access to other adult cardiology subspecialties, including electrophysiology, coronary artery intervention, and heart failure as well as internal medicine, in particular nephrology and hepatology. With the advancing age of the GUCH population, cardiac surgery may need to involve not only congenital heart surgeons but also adult, with joint operating for repair of not only the congenital lesion but repair of valvular heart disease and coronary grafting. Any invasive CHD procedures (surgery, catheter intervention, EP) should be performed under direct consultation with GUCH experts. All GUCH patients considered for CHD surgery should be discussed in multi-disciplinary conferences.

Finally, there should be strong links and referral pathways to a transplant centre with GUCH experience. Non-cardiac surgery can be high risk in patients with complex CHD and collaboration between GUCH specialists and other surgical specialties is essential. Input from orthopaedics is important for the musculoskeletal problems associated with complex CHD but also for the age-related acquired skeletal and joint problems. Additional specialist input may be needed from geneticists, microbiologists, psychologists, social workers, sports medicine experts, and palliative care teams, the latter providing important care and support for the patient and families of those with CHD facing end-of-life issues.

The specialist GUCH centre should ideally be located in an environment which promotes research and innovation, with strong academic and university links.

Table 1 Staff requirements of a specialist grown-up congenital heart centre

Adult/paediatric cardiologist with GUCH certification	At least 2
GUCH imaging specialist (echo, CMR, CT)	At least 2
Congenital invasive cardiologist	At least 2
CHD surgeon	At least 2
Anaesthesiologist with CHD experience and expertise	At least 2
Invasive electrophysiologist with GUCH experience	At least 1
Psychologist	At least 1
Social worker	At least 1
Cardiovascular pathologist	At least 1

Collaboration and infrastructural requirements

Specialist GUCH centres do not only need the above-mentioned services/subspecialties on site but also have to provide coordinated collaborative programmes for clinical service, research, and training. This must include regular conferences, rotation for physicians in training and joint projects (e.g. audit and outcome studies) primarily within GUCH cardiology, adult cardiology, paediatric cardiology, and cardiac surgery/anaesthesia. In addition, collaborations with genetics, gynaecology/obstetrics and radiology must be provided.

In addition to the basic infrastructure of a specialist GUCH centre, which includes dedicated medical staff with nursing and secretarial support, there must be a well-organized outpatient clinic, inpatient facilities and easy access to advanced imaging, electrophysiology, and cardiac catheterization laboratories. There must be a 24-h emergency service for GUCH patients with appropriate on-call arrangements for access to expert GUCH specialist input.

Staff requirements

The specialist GUCH centre must be led by a fully trained GUCH cardiologist with extensive experience and expertise in this field (Table 1). There must be at least one additional GUCH cardiologist with sufficient training and experience to deputize for the lead in their absence to ensure continuity of care at all times. Similarly, there must be at least two specialists in congenital imaging, two interventional cardiologists with GUCH expertise, two CHD surgeons and anaesthetists with CHD experience and at least one interventional electrophysiologist with GUCH experience. Psychosocial services must also be available. Specialist nurses can play an important role in delivering patient care, but opportunities differ in European countries, depending on the healthcare system. If national professional nursing bodies allow such specialization there should be a minimum of two specialist nurses within the GUCH team structure.

Equipment requirements

All diagnostic facilities including ECG, Holter monitoring, echocardiography, cardiopulmonary exercise testing, cardiac magnetic resonance imaging (CMR) and multi-slice computed tomography must be

available and readily accessible (Table 2). Cardiac catheterization (including all interventions) and electrophysiology laboratories (including mapping and ablation systems) must be provided, and there needs to be the facility for pacemaker and ICD implantation with appropriate after-care and follow-up. Fully equipped cardiac theatres and an intensive care unit are mandatory.

Not only physicians but also the technical staff and the lab itself must be prepared to take care of ACHD patients.

Inpatient and outpatient facilities must be adult age appropriate.

Additional requirements

Specialist GUCH centres must provide training programmes for subspecialization in GUCH. They should also participate in quality control programmes, registries, and research and audits of clinical outcome. Scientific activity should be made visible by peer reviewed publications and presentations.

Training in the subspecialty of grown-up congenital heart disease

Despite the fact that many European national cardiology training guidelines recognize the need for specific training in CHD, most cardiologists have little experience or understanding of how to manage GUCH patients.⁶ Although paediatric cardiologists have a great understanding of congenital heart disease, their knowledge of the long-term course in adults and the acquired diseases they may develop with advancing age is limited. In particular, patients who have undergone what are now considered outdated surgical treatments, for example, the atrial switch for transposition of the great arteries and conditions such as Eisenmenger syndrome, are no longer part of current paediatric cardiology practice. Thus, specific formal training in GUCH is essential for both adult and paediatric cardiologists so that they gain the necessary knowledge and experience to provide the best care for these patients. The gaps in knowledge and experience, however, differ between adult and paediatric trained cardiologists and their training needs are different. Initial training recommendations included some training in paediatric cardiology for adult cardiologists and vice versa.^{5,7} It is acknowledged, however, that although such an exchange may be ideal this may often in reality be difficult to facilitate (structural/organizational barriers, payment, clinical duties) and therefore local arrangements, which allow 'training experiences' in paediatric and adult cardiology may also be considered. Daily practice has demonstrated that an appropriate specialist GUCH centre (as described above) may offer an opportunity for adequate training for both paediatric and adult cardiologists. Prolonged time in such a specialist centre may therefore replace a formal 'cross-training'.

Duration of the training

A training period of 24 months is recommended to complete subspecialty training in GUCH. These 24 months should include 18 months in a specialist GUCH centre (requirements see above) and 6 months in general adult cardiology (including CCU, heart failure, arrhythmia, and outpatient clinic) for paediatric cardiology trainees and 6 months of paediatric cardiology (including inpatients and clinic) for adult cardiology trainees. Alternatively, these 6 months could be undertaken

Table 2 Equipment requirements of specialist grown-up congenital heart disease centres

ECG
Holter monitoring
Stress ECG
Ambulatory blood pressure monitoring
Event recorder
Cardiopulmonary exercise testing
Echocardiography (including transoesophageal echo, 3D echo)
CMR imaging
Cardiac computed tomography
Catheterization laboratory
Electrophysiology laboratory
Pacemaker/ICD implantation
Pacemaker/ICD after-care equipment
Cardiac surgery operating room

in a specialist GUCH centre with 'learning experiences' as outlined above.

Training contents

Grown-up congenital heart disease cardiologists must have an understanding of the anatomy and pathophysiology of the different congenital cardiac malformations and the surgical or interventional procedures performed. They must have knowledge of the natural history and long-term course of the various lesions as well as the clinical skill, which enable accurate diagnosis and treatment. An understanding of the optimal timing for intervention in un-operated lesions and when a re-intervention might be needed is of critical importance.

Any GUCH subspecialty training programme must guarantee that trainees see sufficient numbers of GUCH patients across the entire spectrum of CHD complexity, in particular those of moderate and great complexity (see Supplementary material online, *Table S2* and *S3*). Supplementary material online, *Table S4* summarizes general knowledge and experience required for qualification in GUCH. Non-invasive imaging is a key diagnostic tool in CHD, and echocardiography skills in CHD are mandatory. Certification for CHD echocardiography is provided by the European Association of Echocardiography (<http://www.esccardio.org/communities/EAE/accreditation/Pages/welcome.aspx>).

Grown-up congenital heart disease cardiologists should also be able to interpret magnetic resonance imaging and computed tomography in CHD (requirements for the training in ACHD magnetic resonance imaging have been addressed in a separate position paper¹²). Grown-up congenital heart disease cardiologists must be able to interpret invasively derived haemodynamic data and have knowledge of electrophysiology studies and catheter interventions in CHD, without having necessarily performed such procedures themselves. *Table 3* summarizes the performance targets required for certification. The numbers are based on expert consensus.

Since GUCH patients may also develop acquired cardiovascular disease, particularly coronary artery and peripheral artery disease,

valvular heart disease and systemic hypertension, GUCH cardiologists must be familiar with the diagnosis and treatment of these conditions and any preventive treatment strategies.

Grown-up congenital heart disease cardiologists must be competent at risk assessment for pregnancy in CHD and the management of pregnancy, delivery and postpartum care, as well as assessing risk for non-cardiac surgeries and planning peri-operative management.^{13–15} Grown-up congenital heart disease cardiologists must also be familiar with the psychosocial aspects of CHD, recommendations for physical activity (sports, occupation) and other lifestyle issues such as insurance, driver's license, etc.

Certification

Grown-up congenital heart disease experience and training must be recorded in a logbook, which will be verified by an authorized training supervisor in the specialist GUCH centre. A detailed report by the director/lead of the GUCH programme will confirm that all of the requirements (listed in this position paper) for both the GUCH centre itself and the trainee have been fulfilled. When training has been completed, it will be concluded by a formal examination. The writing committee of this position paper recommends that the final exam should be organized and undertaken jointly by the national societies of adult and paediatric cardiology. If national societies agree with these recommendations and can demonstrate adherence with this accreditation process, including potential site visits, external auditing of exams and log-books, then a certification for the subspecialty 'Grown-up congenital heart disease' will be awarded by the ESC.

Supplementary material

Supplementary material is available at *European Heart Journal* online. Funding

Conflict of interest: none declared .

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Table 3 Training in subspecialty grown-up congenital heart disease: specific recommendations

Outpatients [majority with great or moderate complexity (see Supplementary material online, <i>Table S2</i> and <i>S3</i>)]	≥400
Inpatients	≥200
Echocardiography in CHD	
TTE performed	≥250
TEE performed	≥50
CMR/CT in CHD	
Interpreted	≥50
Heart catheterization in CHD	
Interpreted	≥30
Catheter intervention in CHD	
Participated	≥20
Electrophysiology in CHD	
Participated	≥5

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CARDIOVASCULAR FLASHLIGHT

doi:10.1093/eurheartj/eh417

Online publish-ahead-of-print 2 October 2013

Giant aneurysm of the Valsalva sinus associated with multiple coronary artery aneurysms and patent ductus arteriosus

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We report the case of a 42-year-old female referred to our institution for an incidental diagnosis of an aortic aneurysm.

Multislice 64-CT angiography showed a giant aneurysm of the Valsalva sinus, which extended to all three coronary arteries, and multiple aneurysms along the entire length of the coronary arteries (Panels A–C). The Valsalva sinus aneurysm was eccentric, more pronounced near the right coronary cusp (80 mm diameter) and extended to the origin of the right coronary artery (RCA), which had a diameter of 40 mm (Panels D and E) and an excessive tortuosity. The aneurysm also extended to the left main, which had a diameter of 44 mm. This giant aneurysm co-existed with a patent ductus arteriosus (PDA) (Panels B and D).

Surgical repair of the aneurysm and PDA ligation was performed, which resulted in a reduced size of the aortic root and of the origin of the RCA (Panel F).

Aneurysms of the Valsalva sinus can be congenital or acquired and are very rarely associated with coronary malformations. Coronary artery aneurysms (CAA) are very rare anomalies that occur in only 0.2–0.4% of all congenital heart diseases. The association of CAA with aneurysms of the Valsalva sinus and PDA is an extremely rare condition that has not been previously reported. The coexistence of three significant anomalies indicates a congenital aetiology of this complex malformation. The impressive size of the aneurysm and the aspect of the coronary arteries are striking and unique features of this case.

