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Guidelines

# Management of Grown Up Congenital Heart Disease

## The Task Force on the Management of Grown Up Congenital Heart Disease of the European Society of Cardiology

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## Preamble

Guidelines aim to present all the relevant evidence on a particular issue in order to help physicians to weigh the benefits and risks of a particular diagnostic or therapeutic procedure. They should be helpful in everyday clinical decision-making.

A great number of guidelines have been issued in recent years by different organizations-European Society of Cardiology (ESC), American Heart Association (AHA), American College of Cardiology (ACC), and other related societies. By means of links to web sites of National Societies several hundred guidelines are available. This profusion can put at stake the authority and validity of guidelines, which can only be guaranteed if they have

been developed by an unquestionable decision-making process. This is one of the reasons why the ESC and others have issued recommendations for formulating and issuing guidelines, which are quoted as a preamble or appendix in the final reports.

In spite of the fact that standards for issuing good quality guidelines are well defined, recent surveys of guidelines published in peer-reviewed journals between 1985 and 1998 have shown that methodological standards were not complied within the vast majority of cases. It is therefore of great importance that guidelines and recommendations are presented in formats that are easily interpreted. Subsequently, their implementation programmes must also be well conducted. Attempts have been made to determine whether guidelines improve the quality of clinical practice and the utilization of health resources. In addition, the legal implications of medical guidelines have been discussed and examined, resulting in position documents, which have been published by a specific Task Force.

The *Committee for Practice Guidelines (CPG)* supervises and coordinates the preparation of new *Guidelines* and *Expert Consensus Documents* produced by Task Forces, expert groups or consensus panels. The committee is also responsible for the endorsement of these guidelines or statements.

## 1 Introduction and background

As a result of the success of paediatric cardiology and cardiac surgery over the last three decades, there will shortly be more adults than children with congenital heart disease. Prior to the advent of surgery, less than 20% of children born with congenital heart malformations survived to adult life.<sup>1</sup> Now, most deaths from congenital heart disease occur in adults. The 'new population' of patients with congenital heart disease no longer fits within traditional divisions of training and practice, which have separated adult and paediatric cardiology. Adult cardiologists are not equipped to deal with the range and complexity of grown-up patients with congenital heart disease, whereas paediatric cardiologists cannot be expected to manage the many acquired adult diseases in a paediatric medical environment. Up till now, care has been delivered by a number of enthusiastic centres who have managed the complex medical, surgical and psychosocial needs of the grown-up patients with congenital heart disease. In most countries, however, an organized system is not yet in place. This is needed for continued provision of excellence in

clinical care, accumulation of knowledge about the late outcome of management strategies in childhood (with feedback to paediatric practice) as well as for training.

The lack of information regarding numbers, diagnoses and treatment as well as the regular occurrence of avoidable medical problems in this population is testimony to the deficiencies of the current system. The need to reintegrate paediatric and adult cardiac services, and in particular to provide smooth 'transition' for adolescents is clear.

In 1994, the European Society of Cardiology recognized the need for specialized care of this challenging group of patients by establishing a working group for Grown-Up Congenital Heart Disease (GUCH). Since then, both the size, and complexity of the GUCH population in Europe has continued to increase. In 1996, the Canadian Cardiovascular Society commissioned a consensus conference on adult congenital heart disease and its report has been invaluable in representing 'state of the art' principles of management.<sup>2</sup> The American College of Cardiology organized the 32nd Bethesda Conference on care of the adult with congenital disease in 2000<sup>3</sup> and in 1999, the European Society of Cardiology established this Task Force to evaluate provisions for care for grown-ups with congenital heart disease in Europe and to make recommendations for improvement in organization facilities, training, and research. Members of the Task Force include specialists from Europe and North America who were chosen to provide a broad view of novel information, and to developing the recommendations of previous publications.

In the first part of the Task Force report (sections 1–6), the special healthcare needs of grown-ups with congenital heart disease are discussed and common principles of management, including transition from paediatrics, the need for the establishment of specialist centres, models of network care delivery and training of medical and non-medical staff are set out. In the second part (section 7), we have provided structured guides for the management of common lesions. These represent a consensus view of the panellists, and where possible, are evidence-based. They are intended to assist the practicing clinician in a 'user friendly' manner. We recognise that there are many valid different approaches and that more robust clinical research is vital to provide evidence-based recommendations in this emerging field. Nevertheless, we believe that our 'summaries' will be helpful and enable clinicians to access useful information 'at a glance'.

This report is intended to promote collaboration between the various professional groups involved in

the care of adolescents and adults with congenital heart disease, administrators and those who provide resources for health care. Sustained effort to implement the recommendations of this Task Force will be required in order to bring to full fruition the huge successes achieved in the treatment of congenital heart disease in children over the last three decades. The participants humbly acknowledge the many unresolved issues and uncertainties of grown-up congenital heart disease and are all involved in efforts to improve care for this challenging and rewarding group of patients.

### 1.1 Size and composition of the GUCH population

Remarkably few data are currently available on the size and composition of the population of grown-ups with congenital heart disease. Healthcare planning and resource allocation has been based largely on estimates of the incidence of congenital heart disease in infancy, survival through childhood together with the number of 'new' cases such as atrial septal defect, coarctation, Ebstein's anomaly or congenitally corrected transposition which may be diagnosed for the first time in childhood, adolescence and adult life. Such estimates were published by the Bethesda Conference, which highlighted the wide confidence intervals of projected numbers.<sup>3</sup> It has been difficult to collect 'real' numbers because many patients, even those with complex defects attend non-specialized clinics or are completely lost to follow-up.<sup>4</sup> In The Netherlands, for example, it is estimated that there should be approximately 20 000 grown-ups with congenital heart disease but only 8000 are seen in hospital clinics.<sup>5</sup>

There is an increase not only in the size of the population but also in the proportion of patients with complex lesions. Service planning needs to be based on the numbers of grown-ups with congenital heart disease for whom specialist care is essential. Many patients with simpler lesions (e.g. small ventricular septal defect, repaired atrial septal defects, mild pulmonary stenosis, bicuspid aortic valve) can be managed either exclusively in adult general cardiac units or jointly with the specialist unit. In this report, we have stratified care recommendations into three levels (exclusive follow-up in the specialized unit level 1, shared care with local informed adult unit level 2 and predominantly non-specialist care level 3). The proportion of the total population of grown-ups with congenital heart disease and complex lesions requiring either exclusive specialized care or close interaction with a general

adult cardiac clinic (level 1 and 2) has been estimated at 25–50%. A recent survey in the North East Region of the United Kingdom has attempted to predict both the number of patients with congenital heart disease who will survive into adulthood as well as the number who will need specialist care. This used data from a 10-year period (1985–1994) of a congenital heart disease database together with predicted survival of individual lesions from the published literature. When the results were extrapolated to the whole of the United Kingdom, the predicted annual increase in numbers of grown-ups with congenital heart disease was at approximately 1600, with 800 requiring specialized follow-up.<sup>6</sup>

The number of grown-up congenital heart disease patients with individual lesions depends on the incidence at birth, early mortality in childhood as well as the rate of late death. In the absence of hard figures we have developed a simple programme, which enables prediction of late survival rates by entering estimates for each of these outcome determinants. This is available on the ESC website (Appendix 2) and will be useful for planning of resource requirements and funding. A European survey of the numbers of grown-ups with congenital heart disease has been commissioned by the European Society of Cardiology in 24 countries, but few European countries have the necessary established database.

The establishment of specialized centres to manage the complex grown-up heart disease population is a priority. These centres will provide the basis for research into new areas of cardiology, such as the interaction between congenital and acquired heart problems in older patients. Specialized centres should not 'disenfranchise' local physicians, both in general cardiology and primary care who have an important role in a hierarchical local, regional and supraregional service. This report provides a model for such care delivery, which will need to be modified in each of the different health care systems operating in European countries.

## 2 Organization of care

### 2.1 Transition from paediatric to adult care

The arrangement of transition from the paediatric cardiology clinic to the adult service is a particular challenge.<sup>7</sup> 'Adolescence' has no absolute age limits and a degree of flexibility is essential, depending on the intellectual and emotional maturity of the patient as well as other issues such as the presence of coexisting disease.<sup>7</sup> A specialist

transition clinic is highly desirable to minimize anxiety for the patient and their families as well as disruption in care provision. This is the first step to the creation of a successful service for the grown-up population with congenital heart disease and the following recommendations can be made:

- 1 Paediatric cardiologists should begin to inform patients and families regarding transition from around the age of 12, with a flexible policy of transition at age 14–16 years. Subsequent transfer to the adult service can again occur at a flexible age of approximately 18 years. Each paediatric cardiac unit should establish a co-ordinated process to link with a specialist centre for grown-ups with congenital heart disease.
- 2 The patient and their family will have developed a firm bond with the paediatric cardiologist over many years and it is desirable that the paediatric cardiologist is involved in the transition service, together with the adult specialist(s).
- 3 The transition clinic requires input from administration and other healthcare professionals. The nurse specialist is a key person and should be experienced in counselling of adolescents and their families and be responsible for coordinating transfer arrangements.
- 4 The patient and their family should be given a detailed written plan in advance of handover. This should include key information about treatment in childhood (such as previous investigations and operations). It is part of an education process, which must be tailored to the varying levels of maturity and intellect of individual patients. The adolescent and their family need to understand their cardiac condition, healthcare needs and prognosis. In particular, they should know about their medication, possible side effects and interaction with other drugs (including alcohol!) and they should be fully informed about endocarditis prophylaxis. They also require guidance concerning exercise, contraception, pregnancy, career planning, travel and insurance. They must be properly informed of possible future complications of their condition and likely associated symptoms. They must know how to operate within the adult healthcare system in order to obtain appropriate medical advice; both locally and when they are away from home (during studies, occupation and travel). This process must be handled sensitively and cannot occur during a single consultation. The patient should be able to have private discussions, not only with the cardiologist, but also with the nurse specialist. Many children reach adolescence with

little understanding of the implications of their condition, due to parental over protection and lack of direct discussion. Parents often find it difficult to let go of their offspring and may need support to allow the adolescent to become independent. Time should be allocated to discuss sensitive issues such as contraception, family planning, pregnancy, recurrence risk and sport.

- 5 The carefully considered plan of medical management (including follow-up) should be based on the patient's condition and prognosis, education and availability of local medical services. Unit protocols are very useful and the plan needs to be communicated to the primary care physician and other doctors involved (e.g. university healthcare services). Shared care with local physicians is appropriate for many patients. This is particularly important in special circumstances, such as non-cardiac surgery or emergencies. Close liaison and good communication needs to be established at the transition stage.
- 6 Transition of care should be a gradual process both for the patient and the medical practitioners. It is essential not to bombard the patient with an overwhelming amount of information, which can induce denial, and lack of attendance. There should be continued opportunities for joint discussion between paediatric and adult specialists (both medical and surgical) and there should be feedback from both the transition and adult clinics to the paediatric cardiology team.

## 2.2 Network of specialist centres with models of delivery

### 2.2.1 An overview of the current situation

The healthcare needs of many grown-ups with congenital heart disease are not currently being met in Europe and elsewhere. Two fundamental impediments to the improvement of their care are:

- 1 A critical shortage of trained and experienced professionals of the types described below.
- 2 An inadequate number of centres of excellence to lead the national and regional efforts to provide high quality care. A structured hierarchical model of care delivery is required which should be regionalized. The establishment of major full service 'centres' would contrast with the excess of partial service 'centres' currently caring for this population. To be effective, the major centres must have sufficient volumes of patients and procedures to develop and maintain high levels of performance.<sup>8</sup> There is an urgent need of leadership from governments and professional

organizations to work towards such a coordinated system aimed at optimizing patient care.

### 2.2.2 The beginnings of a solution

Given the shortage of staff with training and expertise in the care of grown-ups with congenital heart disease, improvement must start by developing and employing the skilled personnel needed to lead and help coordinate this work in specialist centres. This process would serve as a focus for excellence in clinical care, as well as enabling training and research.

The specialist unit should be located in an adult medical environment with multi-disciplinary speciality provision and be associated with strong paediatric cardiology groups. Indeed, all specialist centres for paediatric cardiology must have defined care pathways for the appropriate transfer of patients to the grown-up congenital heart disease service. Each specialist centre should serve a population of approximately 5–10 million people and they should function within their local medical communities. Cardiologists and primary care physicians should be encouraged to establish a referral relationship with the specialist centres, and this should include provision of timely telephone advice, informal consultation, rapid consultant referrals as well as collaboration in patient follow-up (the specialist centre should include cardiologist(s) with training in management of grown-ups with congenital heart disease in a collaborative team including cardiac surgeons, anaesthetists and intensivists). The cardiologist should be familiar with echocardiography (including transoesophageal echocardiography) and diagnostic cardiac catheterization and at least one per centre should have experience in interventional catheterization. Access to an electrophysiologist with expertise in arrhythmia management in congenital heart disease, pacemaker insertion, ablation and defibrillator implantation is also essential. Specialist imaging including MRI and CT is required. There should be close links with other specialist departments and, in particular, the provision of a joint service with obstetrics to manage high-risk pregnancies. Access to a cardiac pathologist with an interest in congenital cardiac malformations is also highly desirable. A minimum of two congenital heart surgeons (often shared with paediatric cardiology units) is needed, together with the appropriate anaesthetists and intensive care and surgical teams. An association with a transplant centre should also be established.

Specialist nurses are crucial and often provide the 'glue' that connects the various components, as

well as delivering excellence in patient care. All of the unit staff need to work in a dedicated environment with appropriate in-patient and out-patient facilities and a full range of invasive and non-invasive investigational techniques (see below).

## 2.3 Delivery of patient care

Patients should be transferred to the service for grown-up congenital heart disease with a clear management and follow-up plan, with transfer of appropriate information (see above 2.1). All patients should be seen for initial consultation in the specialist adult cardiac services at least once, to establish a relationship with the team as well as to provide familiarity with the new environment. This process will minimize the number of patients who are lost to follow-up during this key period.

Subsequently, patient follow-up care can be stratified in to three levels:

- 1 Patients who require care exclusively in the specialist centre
- 2 Patients in whom shared care can be established with the appropriate general adult cardiac services
- 3 Patients who can be managed in 'non-specialist' clinics (with access to specialized care if required).

These levels are used in the care recommendations for individual lesions in section 7 of this document.

Patients with congenital heart disease who should be seen within the specialist centre include those with the prospect of premature death, re-operation or complications of their condition and those whose condition is unfamiliar to general cardiologists. Any patient who develops a new clinical problem related to their congenital heart defect should be referred for re-evaluation in the specialist unit. Furthermore, consultation should occur with the specialist unit prior to any intervention in a grown-up with congenital heart disease. A telephone call may be all that is required to avoid a disaster during a seemingly innocuous procedure (e.g. non-cardiac surgery in an Eisenmenger patient).

### 2.3.1 Management recommendations

European representatives of the GUCH Working Group of the ESC have had substantial input into the previously published for the management of these patients prior to this report.<sup>2,3</sup> Physicians and patients who are interested in accessing such information may connect to it on [www.achd-library.com](http://www.achd-library.com)

or access the ESC website ([www.escardio.org](http://www.escardio.org)) for the current Task Force recommendations. These provide point-to-point summaries of the major conditions including management recommendations and supporting references. Recruitment and training efforts to build professional teams who will become regional anchors of care should be begun without delay, in order to address the manpower shortage of adequately trained and experienced experts in this field.

### 3 Training of practitioners in grown-up congenital heart disease

Despite the fact that many European national training guidelines for cardiology recognise the need for training in congenital heart disease, most cardiologists have virtually no experience or understanding of management of grown-ups with congenital heart disease. Most units, therefore, rely on an extended role for paediatric cardiologists who work in conjunction with 'interested' adult cardiology colleagues. There is an urgent need to improve this situation by defining and implementing educational requirements for a workforce to staff specialized units for the growing population of adult patients. Appropriate specialist cardiologists may come from trainees in paediatric cardiology or adult cardiology. They should have the following knowledge and skills:

- Expertise of congenital heart malformations and management in infancy and childhood
- Expertise in general medicine and non-cardiac diagnosis in adults
- Expertise in adult cardiology including coronary artery disease management
- Skill in the following procedures in adults with special reference to congenital heart disease:
  - Echocardiography (including transoesophageal)
  - Cardiac catheterization
  - Pacing and electrophysiology
  - Postoperative care
- Understanding of the physiological changes of pregnancy
- Understanding of the psychosocial aspects of adolescence
- Experience of life style counselling for adolescents and adults with congenital heart disease
- Expertise in clinical research methodology

In the specialist centre, there may be advantages to training individuals from both paediatric

and adult backgrounds, to facilitate smooth transition of care for the younger patients as well as provide good care for acquired problems in the older patients.

Efforts are being made in several countries to define training programmes for specialists in the care of grown-ups with congenital heart disease. The following is proposed for trainees from paediatric cardiology:<sup>4</sup>

- Three years in general paediatric cardiology
- 6–12 months in general medicine and adult cardiology
- At least 12–18 months in a specialist grown-up congenital heart disease centre
- For those aiming at an academic career in grown-up congenital heart disease, an additional year in research or specialist training is required

The specific requirements for training are familiarity with:

- 1 Common adult cardiac problems, such as ischaemic heart disease, hypertension, arrhythmias and their treatment
- 2 Coronary angiography (there is no need to learn coronary angioplasty)
- 3 Recognition and management of arrhythmias related to operations for congenital heart disease
- 4 Management of pacemakers and practical experience in their insertion. Management of implanted catheter defibrillators
- 5 Management of middle-aged and elderly patients
- 6 Problems of pregnancy in relation to cardiac problems, and effects of drugs on the patient and foetus
- 7 Understanding the indications and contraindications of various methods of contraception
- 8 Advice about exercise in various congenital heart abnormalities, both un-operated and operated and what limitations, if any, apply
- 9 Advice concerning driving of motor vehicles
- 10 Information about Life insurance and mortgages for patients with various un-operated and operated forms of congenital heart disease
- 11 Vocational advice

For the trainee from the adult cardiological background, training recommendations are:

- Three years in general cardiology including general medicine
- One year of paediatric cardiology in a centre with an active surgical and catheter intervention programme

- At least 12-18 months of training in a specialist GUCH centre for those aiming at an academic career in grown-up congenital heart disease
- Additional year in research or specialist training is required

Specific requirements for training are:

- 1 Foundation courses on congenital heart malformations and echocardiography
- 2 Experience of echocardiography in infants and children, preoperatively and postoperatively. At least 250 echocardiographic examinations should be performed and an additional 25 transoesophageal echocardiograms
- 3 Experience of cardiac catheterization and angiography of common congenital heart anomalies. A minimum of 100 procedures should be performed independently
- 4 Familiarity with interventional procedures in congenital heart disease with participation in at least 25 procedures
- 5 Involvement in management of, and responsibility for at least 300 patients with congenital heart disease. These should include patients in the early postoperative period and those assessed during long-term follow-up
- 6 Attendance at weekly conferences of paediatric cardiologists and cardiac surgeons and participation in all of the teaching activities of the department
- 7 Knowledge of genetic implications and familiarity with genetic counselling
- 8 Understanding of psychosocial problems of adolescence including schooling, bullying, other behavioural issues, such as sex and drugs
- 9 Vocational advice
- 10 Problems of pregnancy in congenital heart disease
- 11 Contraception advice for congenital heart disease

It is envisaged that the specialist in grown-up congenital heart disease will share care with cardiologists in non-specialist centres and for this reason it is recommended that training should be organized for adult cardiologists with 'an interest' in grown-ups with congenital heart disease. These cardiologists, in addition to their normal training (which would have included echocardiography and non-invasive imaging), would:

- Spend the equivalent of 6 months of full-time training in a specialist GUCH centre
- Attend >20 GUCH clinics at the specialist centre over a 2 year period

- Maintain skills by direct association with a specialist centre and participation in CME accredited educational programmes

This would facilitate a flexible interaction between the informed adult cardiologist and the specialist unit, to optimize both patient care and convenience.

It is recommended that a formal certification process should be established by the European board for the speciality of cardiology, indicating that cardiologists (either from paediatric or adult backgrounds) have fulfilled the training requirements in GUCH disease.

The training requirements for surgeons working in the specialist centres also need careful consideration and are currently being considered by a separate committee. The surgeon must have extensive experience in congenital and acquired cardiovascular disorders, before acquiring expertise in the surgery of grown-ups with congenital heart disease. Re-organization and centralization of care for paediatric cardiac surgery is in progress in many countries, in order to ensure a minimum level of surgical activity to develop skills and optimise results.<sup>8</sup>

The establishment of specialist units for grown-ups with congenital heart disease will provide environments for training of all staff and research opportunities. In this small, but growing, subspecialty, collaboration between centres both nationally and internationally is essential. This would enable fellows to rotate between centres within their structured training programme.

Acceptance of clear training pathways and accreditation, as well as provision of the specialist environments for patient care and staff development is the key to a comprehensive clinical service for grown-ups with congenital heart disease.

## 4 Medical issues

### 4.1 Ventricular function

Accurate measurement of ventricular performance is an important part of the preoperative assessment, perioperative management and later follow-up any patient with heart disease. There is still no consensus as to the ideal technique, modality or index to apply to the analysis of left ventricular (LV) function in the biventricular circulation of adults with acquired heart disease. The issues are amplified in those with congenital heart disease. Abnormal ventricular geometry, the effects of previous surgery, extraordinary loading conditions,

chronic hypoxaemia etc, all conspire to make meaningful analysis difficult. Furthermore, right ventricular (RV) dysfunction may be equally or more important in these patients, and similarly may be affected by the supplementary circulatory abnormalities associated with a congenital abnormality. Nonetheless and however flawed, the assessment of functional performance, timing of intervention, and analysis of response is central to the care of these patients.

While this area remains one of the frontiers of the speciality, and many of its aspects investigational, any unit dealing with adult CHD will require subspecialty expertise in quantitative transthoracic and transoesophageal echo Doppler, magnetic resonance imaging, radionuclide perfusion analysis, graded exercise function and invasive haemodynamic assessment. It is likely that there will be considerable overlap with the general adult or paediatric cardiac service, but specific details of testing, and interpretation of results will require appropriate expertise in grown-up congenital heart disease.

#### 4.1.1 Echo-Doppler

Transthoracic echo-windows for parasternal LV short axis function and 'four-chamber' interrogation for RV and LV long axis function and Doppler studies are rarely difficult to obtain. Many of the measures applied to the assessment of systolic and diastolic dysfunction in acquired heart disease are applicable to the 'corrected' biventricular circulation, although with many caveats. For example, LV shortening fraction should be interpreted with caution when there is significant regional incoordination (which is extremely frequent) or residual left-to-right shunting, and Doppler assessment of LV diastolic function must take account of the possible influence of coexisting RV dysfunction in the biventricular circulation. There are more problems when considering the abnormally connected heart. Reduced shortening of the systemic RV may be a physiologic adaptation to increased afterload, and the presence of an intratrial baffle may make Doppler inflow measurements difficult to interpret. Regional incoordination is usual in the systemic ventricle of the post-Fontan 'univentricular' circulation, and diastolic Doppler characteristics must be interpreted with an understanding of the inherently reduced resting preload. The potential of a relatively new modality, tissue Doppler imaging, is large and particularly appropriate to the study of GUCH patients. The demonstration of regional incoordination is intrinsic to the method.

There is no substitute for sequential data. To a large extent, the demonstration of change is more

important than an, apparently, grossly abnormal single measurement. This reinforces the need for a rigid protocol of regular, standardized analysis, with a readily accessible databasing system. This applies equally to all of the techniques described in this section.

#### 4.1.2 Magnetic resonance imaging

Ventricular volume measurements, while more robust than other modalities, are no less immune to the caveats regarding load etc, than shortening indices obtained by echocardiography. Similarly, the role of such measurements in clinical decision-making remains to be demonstrated. Taking the patient after repair of Tetralogy of Fallot as an example, MRI is unsurpassed in its ability to measure RV volumes, image the RV outflow tract and proximal pulmonary arterial tree, and assess volumetrically the degree of pulmonary regurgitation.<sup>9</sup> Similarly intrinsic myocardial performance can be assessed in a way hitherto unexplored by other techniques. Ventricular mass, thickening, vector change, and contractile geometry remain research investigations, with potential clinical applications. As with all of the techniques discussed in this section, sequential data will likely be the most powerful and the non-invasive nature of this method makes this particularly appropriate. It is not too early to say that magnetic resonance imaging is an essential part of any tertiary or quaternary GUCH Unit.

#### 4.1.3 Radionuclide studies

In the ageing population of GUCH patients, an understanding of the indications for, and access to these techniques is required. At a research level, regional abnormalities of myocardial perfusion at a microvascular level are being increasingly recognized.<sup>10</sup> As yet, the direct implications, management, and potential drug modification of these findings remain investigational.

#### 4.1.4 Invasive-studies

Careful evaluation of routine haemodynamic measurements is implicit in all forms of complex congenital heart disease undergoing invasive diagnostic studies. The more detailed assessment of ventricular performance also remains fundamental to the assessment of the natural history of pre- and postoperative disease. Simple angiographic, dimension-based, indices of ventricular performance add little to similar indices measurable by echocardiography or MRI. All such measurements are affected significantly by loading conditions.

The assessment of intrinsic myocardial performance (contractility, diastolic properties etc.) requires more sophisticated analysis. While clearly remaining an investigational tool, conductance catheterization allows characterization of ventricular performance using the elastance model.<sup>11</sup> Continuous pressure-volume analysis during interventions, more accurately allows description of the response to haemodynamic interventions, therapeutic interventions, and assessment before and after surgery.

The adequate assessment of ventricular function and ventriculo-vascular coupling is one of the most important areas of long term follow up of GUCH patients. Appropriate selection of technique will allow more robust analysis of natural and unnatural history, as well as modification by intervention.

## 4.2 Arrhythmia and pacemakers

Arrhythmia is the main reason for the hospitalization of grown-ups with congenital heart disease and is an increasingly frequent cause of morbidity and mortality.<sup>12</sup> Factors that predispose to arrhythmia include the underlying cardiac defect (e.g. atrial isomerism), haemodynamic changes as part of the natural history (e.g. chamber enlargement, myocardial fibrosis), surgical repair and scarring and residual postoperative haemodynamic abnormalities. Arrhythmia, may itself, lead to haemodynamic decline, particularly in patients with very abnormal post operative circuits who are now surviving into adult life. This strong electrical and mechanical connection emphasizes the need for electrophysiological assessment and management to be closely integrated with care of the underlying heart defect. Correction of residual haemodynamic abnormalities may be the most important tool in the treatment of arrhythmia.

Supraventricular arrhythmia is more frequent than ventricular arrhythmia. Sinus node dysfunction is most common after atrial surgery (e.g. Mustard/Senning, Fontan, and atrial septal defect closure) and supraventricular tachycardia (intra-atrial re-entry tachycardia or atrial flutter) is becoming more frequent with longer follow-up.<sup>13–15</sup> Of note, though under emphasized in the medical literature, atrial flutter is a common cause of problems after Tetralogy of Fallot surgery.<sup>16</sup> The highest incidence of ventricular arrhythmia is seen in aortic stenosis and after repair of Tetralogy of Fallot. Patients with the combination of sustained ventricular tachycardia and abnormal haemodynamics are at the highest risk of syncope and sudden death. QRS prolongation has been observed

with right ventricular dilatation and pulmonary regurgitation in Tetralogy of Fallot follow-up, and this may be a useful marker for risk stratification.<sup>17</sup>

Pharmacological treatment of arrhythmia may be limited by haemodynamic side effects, concomitant sinus node dysfunction and by desire for pregnancy. Many standard anti-arrhythmic drugs have proved very disappointing in grown-ups with congenital heart disease and amiodarone is usually the most effective. Side effects, however, are a particular problem in this population.<sup>18</sup>

Catheter ablation and surgical approaches have been increasingly applied.<sup>19</sup> Despite sophisticated current mapping techniques, success rates remain lower than those in structurally normal hearts, largely because arrhythmia circuits are complicated and often multiple.<sup>20</sup> This situation may improve with technological refinements. One of the most challenging groups has been the 'failing' Fontan patients and a combined electrophysiological-surgical revision strategy has met with some success.<sup>21</sup> A similar approach is often needed for Ebstein's anomaly.

Pacing in grown-ups with congenital heart disease is often difficult due to limited, abnormal access to the heart as well as the abnormal cardiac anatomy itself. The right atrial appendage is often absent or distorted and active fixation electrodes are usually required.<sup>22</sup> Furthermore, intracardial shunts and thromboembolic risk may preclude an endocardial approach. A rate responsive system is required and dual chamber pacing is desirable. In the latter, a sophisticated mode switch algorithm should be available because of the high incidence of supraventricular tachycardia and atrial flutter. Anti-tachycardia pacemakers have been disappointing but new algorithms in the current generation of devices may prove more successful for both treatment and prevention of supraventricular arrhythmias.<sup>23</sup>

Implantable cardioverter defibrillator trials in patients with ischaemic heart disease or dilated cardiomyopathy have shown survival benefit in selected subgroups and it is likely that these devices will be used with increasing frequency in patients with congenital heart disease who are considered at risk of sudden death. This has major funding implications and emphasizes the importance of identification of patients at high risk of malignant arrhythmia and sudden death.

The need for risk stratification, understanding of anatomy and function, choice of drug, catheter/implantable defibrillator or surgical intervention emphasizes the importance of very close integration of the electro-physiologist with the GUCH

cardiologist and surgeon in the specialized team. It should be appreciated, however, that electrophysiological experts with the particular skills required for patients with congenital heart disease are rare and both training and resources need to be increased.

### 4.3 Cyanosis in the GUCH patient

Right to left shunts and the resulting hypoxaemia have profound haematological consequences, which affect many organs.

#### 4.3.1 Haematologic problems

The increase in red cell mass, which accompanies cyanosis, is a compensatory response to improve oxygen transport. The white cell count is usually normal, and the platelet count may be normal or, more often, reduced. The increased red cell mass and consequent increased viscosity increases the risk of stroke, though in adults this is only slightly raised.<sup>24</sup> Most patients have a compensated erythropoiesis with stable haemoglobin that requires no intervention. Therapeutic phlebotomy, therefore, is usually unnecessary unless the haemoglobin is >20 g/dL and the haematocrit is >65%. At these levels of increased red cells, patients often experience symptoms of the 'hyperviscosity syndrome' primarily consisting of headache and poor concentration. These symptoms may be relieved by removal of one unit of blood, *always* with an equal volume replacement of dextrose or saline. Therapeutic phlebotomy, however, is a 'two-edged sword' since erythropoietin may stimulate the bone marrow to produce more red cells. It is recommended, therefore, that therapeutic phlebotomy be performed no more than 2–3 times per year. Repeated phlebotomy depletes the iron stores and may result in the production of iron-deficient red cells. These iron-deficient microcytes are less deformable than iron-replete red cells and increase the risk of stroke by increasing blood viscosity.

The treatment of iron deficiency in a patient with destabilized erythropoiesis is challenging since oral iron frequently results in a rapid and dramatic increase in red cell mass. Administration of one tablet of ferrous sulfate (or gluconate) is recommended with a recheck of the haemoglobin in 7–10 days. The iron should be discontinued if there is a dramatic increase in red cell count.

#### 4.3.2 Haemostasis

Reduced platelet count and abnormal platelet function together with clotting factor deficiencies combine to produce a bleeding tendency in

cyanotic patients, either spontaneously or perioperatively. Gingival bleeding, menorrhagia, and pulmonary hemorrhage (manifesting as haemoptysis) are common. The latter is sometimes fatal. For these reasons, the use of anticoagulants and antiplatelet agents should be confined to well-defined indications, with careful monitoring of the degree of anticoagulation. When the hematocrit is >60%, the citrate concentration in the coagulation tests needs to be adjusted; otherwise the results may be inaccurate.

#### 4.3.3 Renal function

In chronic cyanosis, the renal glomeruli are markedly abnormal and are frequently hypercellular and congested.<sup>25</sup> This results in a reduction of the glomerular filtration rate and increased creatinine levels. Proteinuria is common. Abnormal urate clearance frequently results and this, in addition to the increased turnover of red cells, leads to hyperuricemia and sometimes frank gout. Hyperuricemia without gout is usually well tolerated, however, and does not require intervention.<sup>26</sup>

#### 4.3.4 Gallstones

Bilirubin may be produced from the breakdown of haeme in chronic cyanosis, and calcium bilirubinate gallstones are common in this adult population.<sup>27</sup>

#### 4.3.5 Orthopaedic complications

Hypertrophic osteoarthropathy with thickened, irregular periosteum occurs in adults. This is sometimes accompanied by aching and tenderness especially in the long bones of the legs. Scoliosis is another important complication, which at times may be sufficiently severe to compromise pulmonary function.

#### 4.3.6 Skin

Acne on the face and trunk frequently accompanies cyanosis. It is not just a cosmetic concern as it is a potential source of sepsis and endocarditis.

### 4.4 Pulmonary vascular disease (Fig. 3)

In the last 20 years, early diagnosis and improved infant cardiac surgery have reduced the number of adolescents and adults with pulmonary vascular disease. However, a considerable number of such patients still attend clinics for grown-ups with congenital heart disease. Despite the fact that their underlying condition is irreversible and progressive, there is a considerable return in terms of morbidity and mortality from careful management.

Many patients with Eisenmenger syndrome maintain a good quality of life into early adulthood but subsequently the reduction of effort tolerance and increased cyanosis is usually progressive.<sup>28</sup> The most important clinical events are pulmonary and cerebral complications. Haemoptysis may be due to both pulmonary infarction or to ruptured blood vessels. As in other cyanotic patients, Eisenmenger patients are vulnerable to spontaneous bleeding (e.g. gums, gastric mucosal as well as haemoptysis) and have abnormal haemostasis including prolongation of prothrombitime, APTT and abnormal platelet function. The decision to anticoagulate such patients is, therefore, difficult and controversial. While patients with primary pulmonary hypertension have demonstrated improved survival with anticoagulation, no data exists for Eisenmenger patients and the risks may outweigh any potential benefits. Haemoptysis may be precipitated by stress/excitement or a chest infection and can be copious and fatal. It remains difficult to identify patients with particular risk of pulmonary thromboembolic complications. Similarly, cerebral complications (stroke or abscess) are not necessarily associated with higher haematocrit levels. With good management, patients may survive well into adult life. In a large European collaborative study, the mean age of death of patients with Eisenmenger syndrome and simple congenital heart disease was 32.5 years compared to 25.8 years in those with complex defects.<sup>28</sup> Right ventricular function and age at clinical deterioration were additional prognostic determinants. Eisenmenger patients are at particular risk from pregnancy, dehydration, cardiac and non-cardiac surgery, general anaesthesia, anaemia, chest infections, altitude, intravenous lines and vasodilating drugs. There is approximately a 50% maternal fatality in conjunction with pregnancy and death often occurs after delivery.<sup>24</sup> Pregnancy is therefore contraindicated, even in those with 'moderate' pulmonary hypertension. Other, seemingly innocuous, non-cardiac procedures are also risks factor for death.<sup>24</sup> Careful consideration should therefore be given to the indications for any intervention and procedures should only be carried out in specialist centres with experienced cardiac anaesthetists. A general policy of 'non-intervention', unless absolutely indicated, is recommended to avoid destabilizing the 'balanced physiology'.

A number of the treatments have shown haemodynamic improvement, and in some cases, outcome benefits in primary pulmonary hypertension and may also be applicable in grown-ups with congenital heart disease and Eisenmenger Syndrome.

Systemic vasodilators such as calcium channel blockers should however be given with caution. Prostacycline, endothelin antagonists and Sildenafil may have a role, but no convincing clinical trial evidence is yet available. Long-term oxygen therapy at home for a minimum of 12–15 h per day may improve symptoms significantly but does not modify survival. Ultimately, lung transplantation needs to be considered for some patients.<sup>29</sup>

#### 4.5 Infective endocarditis

The ESC has its own Task Force on infective endocarditis and this document should be referred to for more extensive information.<sup>30</sup>

Most grown-ups with congenital heart disease, but not all, have a life-long risk of infective endocarditis. Education of the patient and their physicians about the risks and importance of early diagnosis needs constant reinforcement. There may be a variety of portals of entry of infection, in addition to dental and surgical procedures. These include body piercing, acne and tattooing. Interventional cardiac catheterization is infrequently associated with endocarditis, but antibiotic prophylaxis is usually given. Similarly, insertion of intrauterine contraceptive devices should be covered and antibiotics are generally recommended at childbirth.

Delay in diagnosis and referral is common and antibiotics are frequently prescribed before the diagnosis of endocarditis is considered or blood cultures are taken. Two blood cultures are adequate for microbiological isolation in approximately 95% of patients with the usual organisms who have not received antibiotics, but more should be performed to exclude clinically suspected infected endocarditis. It is easy to miss vegetations in adults using transthoracic echocardiography, particularly when this is performed by operators inexperienced in the investigation of congenital heart disease. The use of transoesophageal echocardiography increases the detection of vegetations considerably.

Prompt referral to the specialist unit is usually indicated as haemodynamic deterioration may be rapid and surgical management may be required. Infective endocarditis accounted for approximately 4% of admissions to a specialized unit for grown-ups with congenital heart disease in the UK.<sup>4</sup>

Not all patients are at risk and antibiotic prophylaxis recommendations vary. Infected endocarditis is not reported in secundum atrial septal defect, totally anomalous pulmonary venous connection

and is extremely rare after the closure of ventricular septal defect, in pulmonary valve stenosis or small patent ductus arteriosus. Prophylaxis policies after interventional catheterization procedures involving device implantation vary, and are not 'evidence based'. Antibiotic treatment and overall patient management should be undertaken in collaboration with an infectious disease specialist. Fungal endocarditis may be particularly difficult to treat and other unusual organisms may be responsible. The cardiac surgeon needs to be involved early, as surgical replacement of infected prosthetic material may be required. The ESC and AHA have published specific antibiotic regimens.<sup>30,31</sup>

#### 4.6 Imaging in adults with congenital cardiac disease

There has been a shift from cardiac catheterization to non-invasive imaging modalities for patients with grown-up congenital heart disease, which is similar to that which has occurred in paediatric cardiology over the last two decades. These include echocardiography (transthoracic and transoesophageal) and MRI, which are essential in specialist GUCH centres. Cardiac catheterization is now reserved for resolution of specific anatomical or physiological questions (e.g. coronary arteries, pulmonary vascular resistance) or for intervention (see below). Transthoracic echo in adults is less effective than in children at describing complex intracardiac and extracardiac anatomy and physiology. Transoesophageal echocardiography often provides definitive information but has the disadvantage of usually requiring sedation or anaesthesia. MRI is increasingly becoming the investigation of choice for grown-ups with congenital heart disease providing 3D reconstruction of anatomy, improved temporal and spatial resolution as well as physiological information (Fig. 1). It has proved particularly useful for evaluation of right ventricular volume and mass, right ventricle to pulmonary artery conduit/valve function as well as for the study of pulmonary arteries, coarctation and systemic and pulmonary venous anomalies.<sup>32,33</sup> Interventional procedures such as balloon dilatation and radio frequency ablation can now be carried out using MRI/catheter fluoroscopy techniques, further extending the role of this imaging modality. Until recently, computed tomography (CT) played only a minor role in the evaluation of congenital heart disease. Newer technologies however, such as ultra fast CT and spiral CT have reduced scanning time and provide excellent imaging which can be seen as complementary to echo cardiography and MRI.<sup>34</sup> All of these imag-

ing techniques require staff with expertise in complex congenital heart disease, in order to avoid the high rates of diagnostic errors reported in routine adult laboratories.<sup>35</sup> This has planning, training and funding implications.

#### 4.7 Interventional catheterization

The rapid increase in the type and application of interventional techniques in congenital heart disease of childhood has been mirrored in the adult population. With the exception of calcified valves, balloon dilatation of congenitally stenotic aortic, pulmonary and rheumatic mitral valves, is relatively successful.

Many of the techniques developed in children were contemporaneously, or subsequently adapted to the treatment of grown-ups with congenital heart disease. Relatively few techniques have become unequivocally established as first choice over their surgical counterparts. The decision to perform an intervention should therefore undergo a similar process of peer-review, and multidisciplinary discussion as with surgery. Randomized studies against the surgical alternative are almost non-existent; registry data are limited, and local series are potentially affected by selection and reporting bias. Nonetheless, an interventional programme is central to any GUCH unit and at least one member of the team should be trained in interventional cardiology. Previous interventional experience in children is probably more relevant than previous experience in coronary intervention, but increasingly, specific experience in GUCH intervention is most desirable. The same can be said for allied medical support (anaesthesia, echocardiography etc.) and technical support within the cardiac catheter laboratory. The laboratory should be equipped with biplane screening and storage equipment, should be large enough to allow simultaneous transoesophageal echocardiography, and be equipped with appropriate catheters, wires and devices.

##### 4.7.1 Techniques

###### 4.7.1.1 Balloon dilation

Balloon dilation of congenitally stenotic aortic, pulmonary and rheumatic mitral valves has been very successful. Aortic valvoplasty is usually performed using a single balloon technique. As in childhood, residual aortic incompetence is the most important complication. A balloon to aortic ratio of <1:1, antegrade dilatation via transeptal puncture, and adenosine cardioplegia have all been used to reduce the incidence of valve damage.<sup>36</sup> Single,



Fig. 1 Magnetic resonance angiography image of severe aortic coarctation in an adult. See section 4.6 – Imaging.

double, or Inoue balloon dilatation of pulmonary valve stenosis is usually successful, but a balloon:annulus ratio of <1.1:1 should be used to avoid pulmonary arterial damage. The role of balloon dilatation of native or postoperative aortic coarctation remains controversial.<sup>37</sup> Dilatation of previous patch aortoplasty carries the highest risk of aortic rupture and should be performed with surgical standby. Balloon dilatation of native aortic disease can often be achieved with excellent results, although failure to relieve stenosis (10–20%) aneurysm formation (5–10%) and restenosis (5–10%) have all been reported.<sup>38</sup> For this reason, there has been an increase in vogue towards balloon dilatation with simultaneous stent implantation.

**4.7.1.2 Balloon dilatation with stent implantation**  
This has become the technique of first choice for branch and distal pulmonary arterial stenosis.<sup>39</sup> Balloon dilatation of right ventricular outflow tract obstruction, and stenotic right ventricular to pulmonary arterial conduits is less widely accepted. Stent implantation also has a role in the treatment of resistant stenoses in surgical venous pathways<sup>40</sup> (Mustard/Senning/Fontan). Systemic arterial stent implantation is more controversial. The utility of stent implantation for coarctation of the aorta, stenotic aorto pulmonary collaterals in complex pulmonary atresia, and stenotic systemic to pul-

monary shunts, while reported, remains to be proven. The need for anticoagulation after stent implantation is not known. Most would fully anticoagulate patients after pulmonary and systemic venous implantation and in those with erythrocytosis.

#### 4.7.1.3 Embolization and occlusion techniques

Unwanted, usually acquired, venous and arterial collaterals and fistulous communications are readily occluded using coil embolization techniques. Similarly, the small arterial duct in adults can be closed using detachable coils, although in larger lesions (4–16 mm), a detachable plug device is required (Fig. 4).

There are several devices available for closure of secundum atrial septal defect and patent foramen ovale. Transcatheter closure of atrial septal defect has become standard practice in most congenital units, for selected cases. Depending on the device, defects of up to 40 mm can be closed, providing there is suitable atrial and septal anatomy. Many of the unanswered questions pertaining to surgical closure of ASD, are relevant to transcatheter closure. The degree of acceptable pulmonary hypertension, the need for concomitant atrial antiarrhythmia surgery and likelihood of symptomatic response, which depends on age at closure are yet to be resolved. Closure of patent foramen ovale in



Fig. 2 Unsightly scars from multiple cardiac procedures that can cause psychosocial problems. See Section 6 – Psychosocial Issues.

patients with early onset, cryptogenic, transient ischaemia attack (TIA) or cerebrovascular accident (CVA) can be achieved with extremely low morbidity, but with little evidence base.<sup>41</sup> Recent trials have suggested the importance of aspirin in determining the level of risk of CVA in the presence of patent foramen ovale.<sup>42</sup> Treatment should be individualized and decision-making part of a multidisciplinary process, which includes a specialist neurologist.

Many of these same 'ASD' devices can be used for occlusion of other unwanted intra and extracardiac communications. Transcatheter closure of congenital ventricular septal defect is rarely indicated, but some ischaemic VSD's may be amenable to closure. Baffle leaks, systemic arterial, coronary, and venous fistulas communications have all been closed using these devices.

#### 4.7.1.4 Percutaneous valve implantation

It is now possible to insert into the right ventricular outflow, a stent mounted bovine jugular venous valve, via a cardiac catheter.<sup>43</sup> Clinical implantation has already been performed and offers the exciting prospect of non-surgical management of



Fig. 3 Chest radiograph of a patient with Eisenmenger's syndrome. See Section 4.4 – Pulmonary Vascular Disease.

pulmonary regurgitation after repair of Tetralogy of Fallot and related conditions. Further work is required to develop a valve, which is suitable for the dilated outflow tract found in many adult patients. An implantable valve that can be used in the systemic circulation also appears possible.

## 4.8 Pregnancy and GUCH

Most grown-ups with congenital heart disease can tolerate a pregnancy with proper care.<sup>44,45</sup> Pre-pregnancy counselling and evaluation is mandatory and should include a physical examination, assessment of haemodynamic status (usually including echocardiography) and functional capacity. Exercise testing may be particularly helpful in this regard. A complete family history should also be taken to offer informed genetic counselling. A review of medications is necessary to avoid any drugs that may be deleterious to the foetus (e.g. ACE inhibitors). The risks to the mother of morbidity and mortality and, when appropriate, the impact of pregnancy on long-term survival should be discussed. In addition, the risks to the foetus of inheriting congenital heart disease should be considered. Patients can be stratified into high, medium and low risk. The highest maternal risk is associated with Eisenmenger syndrome, with a maternal mortality of  $\geq 50\%$  (often after delivery).<sup>46</sup>



Fig. 4 Transcatheter closure of patent arterial duct using an amplatzer device. See Section 4.7 – Interventional Catheterization.

The number of mothers with complex post operative circulations (such as Fontan or Mustard/Senning) remains small and risk stratification is therefore difficult.<sup>47,48</sup> High-risk patients include those with:

- Significant aortic stenosis (mean gradient >40 mmHg, valve area <0.7 cm<sup>2</sup>)
- Significant coarctation, particularly with aortopathy
- Significant mitral stenosis
- Reduced systemic ventricular function
- Mechanical prosthetic valve
- Pulmonary hypertension
- Marfan's syndrome
- Cyanotic heart disease

Appropriate medical management throughout pregnancy for high-risk patients may include:

- Iron and prenatal vitamins to reduce anaemia.
- Monitoring maternal and foetal well-being.
- Foetal cardiac ultrasound, which should be offered to the mother to evaluate potential foetal cardiovascular abnormalities.
- For very high-risk patients, bed rest may be necessary as pregnancy advances with delivery at a tertiary care centre.

- For most patients, vaginal delivery is preferable to caesarean section with few exceptions, unless there are obstetric indications.
- Cardiac monitoring during delivery is appropriate in those with 'fragile' haemodynamic status.

The management of patients with mechanical valves is particularly challenging, particularly those with mitral tilting disc prostheses.<sup>49,50</sup> Heparin treatment may be associated with thromboembolic complications, whether administered subcutaneously or intravenously. The role of low molecular weight heparin is not established. Warfarin treatment may be safer for the mother, but can be associated with foetal embryopathy. This risk appears to be small if the Warfarin dose is <5 mg per day.<sup>51</sup>

Cyanotic heart disease also poses a significant risk for the foetus and this is proportional to the degree of maternal hypoxia. Low birth weight for gestational age and prematurity are common with maternal cyanosis, and for those mothers with an arterial oxygen saturation ≤85%, only 12% of pregnancies are successful.<sup>52</sup> Patients who should be counselled against pregnancy include those with:

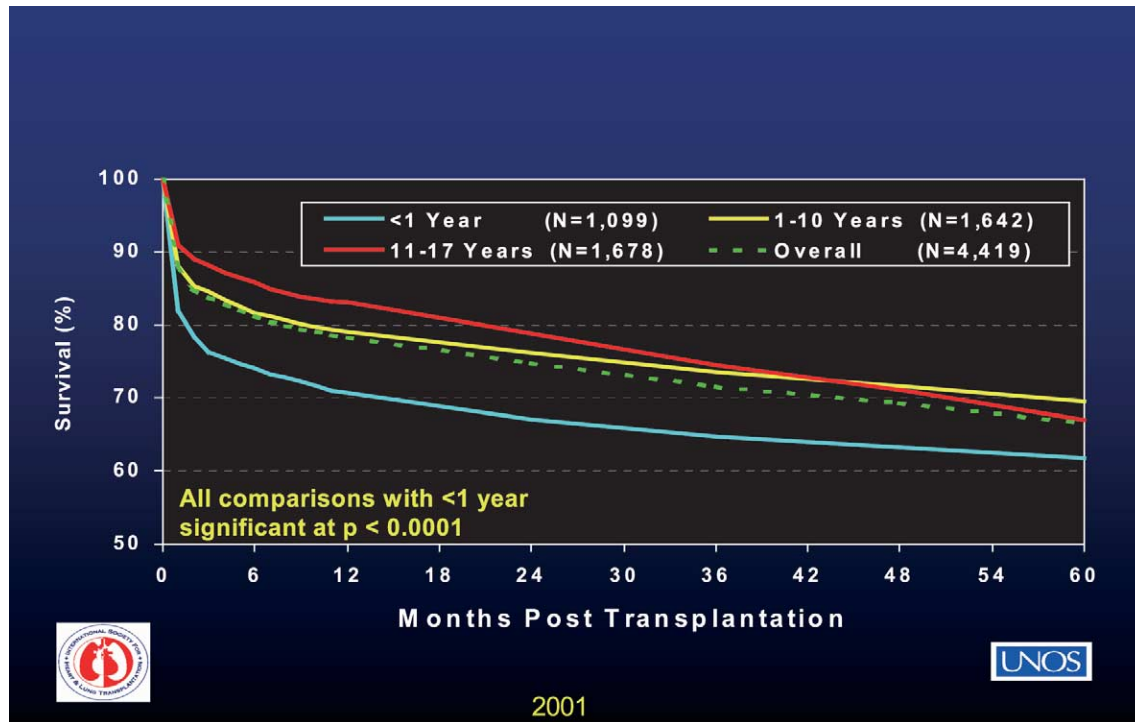


Fig. 5 International Society for Heart & Lung Transplantation results 1982–2000. Congenital Heart Disease comprised 25% of heart transplants in 11–17 year olds group and was a risk factor for survival (OR 1.59). Results in adults with congenital heart disease have not been reported separately. See Section 5.4 – Transplantation.

- Eisenmenger syndrome (50% maternal mortality). Termination of pregnancy is safer, preferably with cardiac anaesthesia.
- Marfan's syndrome with dilated aortic root.
- Severe aortic stenosis/coarctation.
- Systemic ventricular ejection fraction <35%.

Women with congenital heart disease considered at high risk should be managed in a specialized unit, which includes an obstetrician, a cardiologist expert in grown-up congenital heart disease, an anaesthetist and a paediatrician. This team should be involved from early pregnancy and plan monitoring of the pregnancy, mode of delivery and post delivery care. This specialized team can also provide a consultation service for obstetricians and physicians managing 'lower' risk women in other centres.

#### 4.9 Genetic counselling and contraception in GUCH

The risks of pregnancy vary greatly in congenital heart disease and must be weighed against the risks of contraception. Systematic studies, however, on contraception in women with congenital heart disease are lacking.<sup>52</sup> Patient compliance and sexual behaviour must be considered, as they will affect

contraceptive efficacy and the incidence of complications. Usually, the patient chooses her own preferred method, but the physician should provide informed advice. Often, there is no ideal method and the least hazardous one is indicated.

Barrier methods are safe from a cardiovascular standpoint and have a high degree of contraceptive efficacy in compliant couples and women >35 years. Low dose oestrogen combined oral contraceptive pills are very efficacious, but their thrombotic properties may make them hazardous in certain situations, such as after the Fontan operation or in patients with atrial fibrillation/flutter. Combined oral contraceptives are contraindicated in patients at risk of paradoxical embolism, unless they are also receiving anticoagulants. The oestrogen containing contraceptive pill should not be used in patients with pulmonary or systemic hypertension. Medroxyprogesterone injection (Depo-Provera<sup>®</sup>), subcutaneous deposition of levonorgestrel (Norplant<sup>®</sup>) or progesterone only pills are effective, but may cause fluid retention and should not be used in patients with heart failure. Depression and breakthrough bleeding may prevent the use of progesterone pills and there is a higher failure rate than with the combined oral contraceptives.<sup>53</sup>

Intrauterine devices have been associated with an increased incidence of pelvic inflammatory disease and it has been suggested that they carry a risk of endocarditis, especially in cyanotic women, those with artificial valves/shunts/conduits and those with previous endocarditis. However, the reported number of women with intrauterine devices complicated by endocarditis is very low and concerns mainly old-fashioned intrauterine devices. The levonorgestrel-releasing intrauterine device (Levonaova<sup>®</sup>) has a low incidence of pelvic inflammatory disease and a contraceptive efficacy equivalent with that of combined oral contraceptives.<sup>54</sup> It has been approved for use in women at risk of endocarditis. If used in that setting, antibiotic prophylaxis should be given at insertion as well as at extraction. The local release of progesterone reduces the bleeding problems encountered with other intrauterine devices. In women who have not experienced pregnancy, intrauterine devices are not a first choice.

Surgical sterilization may be considered in a woman at high risk from pregnancy, but should not be undertaken without discussion of potential medical advances, which might later allow pregnancy at an acceptable risk.

#### 4.9.1 Recurrence risk/genetic counselling

There is extensive information about recurrence risk of congenital heart disease in siblings but until recently, few data about recurrence risk in couples where the mother or father has a congenital heart defect. The recurrence rate of congenital heart disease in offspring ranges from to 2–50% and there is a higher risk when the mother, rather than the father, has cardiac disease.<sup>55–57</sup> The highest recurrence risks are in single gene disorders and/or chromosomal abnormalities, such as Marfan's syndrome, Noonan's syndrome and Holt-Oram syndrome. Genetic counselling should be available in the specialized units for grown-ups with congenital heart disease and specific genetic testing is likely to evolve as understanding of the genetic basis of congenital heart disease improves. The specialist unit should also offer foetal echocardiography at 16–18 weeks gestation and chorionic villous sampling or amniocentesis may also be indicated in selected cases. The potential for drugs to affect the foetus should always be considered. In particular, angiotensin converting enzyme inhibitors and angiotensin II receptor blockers should not be used in pregnancy and withdrawal of amiodarone and Warfarin should be considered. Protocols for stopping Warfarin and managing anticoagulation are required.

## 4.10 Comorbidity and syndromes

Congenital and acquired comorbidity is common in grown-ups with congenital heart disease and has an important effect on outcome and treatment. Cognitive and intellectual impairment may be a feature of co-existing heritable or chromosomal syndromes, which are present in 15–20% of congenital heart disease. Such patients are surviving into adult life with increasing frequency due to a more active approach to their treatment in childhood. Alternatively, late problems may result from neurological complications in the perinatal and perioperative period and are likely to pose a serious burden on the families and to produce an important demand on medical and social institutions. This has not yet been addressed or funded in most European countries. Knowledge of the individual characteristics of such syndromes assists cardiac diagnosis and provides a clue to the presence of extra cardiac problems.

Skeletal deformities are common in cyanotic states, especially when a lateral thoracotomy incision was performed in early life. There may also be visible chest or breast deformities, which have psychosocial impact (Fig. 2).

Chronic cyanosis is associated with pulmonary, metabolic and haematological problems (see section 4.3) and sequelae from previous operations or catheterization may include arterial or venous peripheral occlusions and scars on the chest.

Management of comorbidity in grown-ups with congenital heart disease deserves a thorough understanding of pathophysiology by the cardiology team as well as close communication with the various non-cardiac specialists involved (Table 1).

Acquired heart disease will occur with increasing frequency as the population with congenital heart disease ages. Coronary artery disease and/or systemic hypertension can affect the haemodynamics of the congenital heart disease, requiring treatment of both the congenital and acquired disorders. This emphasizes the importance of the close interaction between the specialist unit for grown-ups with congenital heart disease and members of the general adult cardiology department.

## 4.11 Emergencies – adults with congenital cardiac disease

Acute cardiac and non-cardiac emergencies occur in grown-ups with congenital heart disease and their optimal management may be above the ability of emergency room, general medical or adult cardiology staff.<sup>58</sup> The most frequent are arrhythmia,

**Table 1** Syndromes associated with mental deficits

Syndrome	General features	Cardiac defect
Alcohol Syndrome	Facial and growth anomaly, mental retardation	ASD, VSD (30%)
Down (trisomy 21)	Mental retardation, typical facies, lymphedema	AVSD (VSD, aortic valve anomaly) in 40%
Noonan	Turner like phenotype, normal chromosomes, mental retardation	Coarctation, HCM, ASD, PS
Turner	Chromosome XO, skeletal and mental deficits	Coarctation in 35%, ±bicuspid aortic valve
Williams Beuren	Facial dysplasia, hypervitaminosis D, hypocalcaemia, mental retardation	Supravalvar aortic stenosis, sometimes with multiple pulmonary artery stenoses

infections, heart failure, cerebral ischaemia or aortic root problems. It may be possible to provide appropriate initial treatment in the local hospital (usually after consultation with the specialist centre) but patients with more severe complications or more complex congenital heart disease will usually require transfer to the specialist centre. Geographical planning of the hierarchical care delivery model is thus very important. At present, the limited available expertise has developed in an unplanned fashion and long distances between specialist centres may delay diagnosis and treatment and actually discourage appropriate transfer.

## 5 Surgical issues

### 5.1 Cardiac surgery

Adolescents and adults with congenital heart disease who require surgery fall into three categories: (1) those who have not previously undergone operation, (2) those who have had palliative surgery and (3) those who have had reparative surgery. In each category, there are several considerations, which make surgery, in this population, different from other types of cardiac surgery (either adult surgery for acquired heart disease or paediatric surgery for congenital defects). There are strong arguments for concentrating surgical management of grown-ups with congenital heart disease into specialist units for both care provision and training. Surgery can be performed safely only by teams who have extensive experience in the management of congenital heart defects in infants and children as well as knowledge of the principles of conventional adult cardiac surgery.

#### 5.1.1 General planning of the operation

The first operation may be required in patients with simple defects (e.g. atrial septal defect) but more often is needed in patients with complex malfor-

mations (e.g. pulmonary atresia with ventricular septal defect). In patients who have previously undergone palliative or reparative surgery, the consequences of previous operations may add to the complexity of the management of the primary defect. Thorough cardiac evaluation is mandatory in all cases. Surgical planning requires knowledge of the basic congenital malformation, of the previous surgical procedures and of the potential residual or recurrent lesions after these operations. The surgical team needs to be intimately involved in the review of the diagnostic information, the decision-making and planning of the patients' overall management. It is mandatory that the previous surgical reports be available.

In adult cardiac surgery for acquired heart disease, the risk and benefits of most surgical procedures are well established. In contrast, every grown-up with congenital heart disease poses specific problems and the risk/benefit ratio of any proposed surgical procedure is often difficult to assess. It may therefore be difficult to communicate the necessary information about risks and benefits to the patient.

#### 5.1.2 Specific surgical challenges

Some surgical problems apply to all first operations or re-operations in adults with congenital heart disease.

##### 5.1.2.1 Preservation of myocardial function

In most patients, preoperative ventricular function is abnormal as a result of ventricular morphology, ventricular hypertrophy and long-standing pressure or volume overload. Myocardial fibrosis and ischaemia as well as the sequelae of previous operations may also influence ventricular function and require meticulous preoperative assessment.

To achieve optimal intraoperative preservation of myocardial function, some recommendations can be made:

- 1 Aortic cross clamping should be avoided where possible. Most operations involving the right heart (e.g. replacement of right ventricle to pulmonary arteryvalved conduit or extracardiac Fontan procedure) can be carried out under normothermic cardiopulmonary bypass with mono or bicaval cannulation and a beating heart.
- 2 When aortic cross clamping is necessary, cross-clamp time should be kept as short as possible and particular attention should be paid to cardioplegic myocardial preservation.

Strategies for myocardial protection and cardioplegia should include:

- a Use of the appropriate cardioplegic solution.
- b Induction of myocardial hypothermia using a cold blood cardioplegic solution.
- c Maintenance of diastolic arrest and hypothermia using multi-dose blood cardioplegia. This is particularly important in cyanotic adults, in whom non-coronary collateral vessels to the heart may result in wash out of cardioplegia and myocardial re-warming.
- d Enhanced warm blood reperfusion administered prior to aortic unclamping under careful pressure monitoring.
- e Adequate venting of the heart to avoid ventricular distention, wall tension increase and subsequent inadequate delivery of cardioplegic solution.

### 5.1.3 Blood salvage techniques

Autologous transfusion should be encouraged and be used wherever possible. In cyanotic patients, preoperative phlebotomy may be indicated to improve haemostatic status and the blood should be kept for autologous transfusion. Patients undergoing re-operation after initial reparative surgery are less likely to be cyanotic but they can usually predonate an adequate amount of blood and fresh frozen plasma. Iron repletion is indicated in most cases managed in this way. Erythropoietin administration may allow rapid and timely increases in haematocrit. Patients with congenital heart disease are prone to intraoperative and postoperative bleeding because of relatively long suture lines, an increase in tissue vascularity, intrinsic haemostatic defects and prolonged periods of cardiopulmonary bypass. Replacement of consumed or inactivated clotting factors, particularly platelets, is an important requirement. Aprotinin has proved effective in reducing intraoperative bleeding, particularly in patients undergoing re-operation. The routine intraoperative use of a cell saver system is important. Ultrafiltration (either conventional during cardio-

pulmonary bypass or modified after cessation of bypass) has been shown to result in a significant reduction in the need for postoperative blood transfusion as well as to improve myocardial and lung function and extraction of soluble inflammatory mediators.

### 5.1.4 Redo sternotomy incision

Reopening a sternal incision remains a crucial step, particularly when an enlarged right ventricle or an extracardiac conduit may be apposed or adherent to the back of the sternum, or when the aorta lies anteriorly (transposition of the great arteries). If particular difficulty is anticipated, cannulation of the femoral vessels, institution of bypass and decompression of the heart before reopening the sternum are prudent precautions. When re-operation is anticipated, at previous operation, implantation of a retrosternal prosthetic membrane greatly facilitates reopening of the sternum, although this sometimes makes more difficult the subsequent identification of the mediastinal and cardiac structures.

### 5.1.5 Pulmonary vascular bed abnormalities

In most patients undergoing surgery for acquired heart disease, the pulmonary vascular bed is normal. This is not the case in many adults with congenital heart disease and management of elevated pulmonary vascular resistance is particularly important in the early post operative period. Severe distortion of the pulmonary arteries may be present in previously operated patients (pulmonary artery banding, aortopulmonary shunt or previous surgery involving the pulmonary arteries). Preoperative dilatation and stent implantation is sometimes indicated. Alternatively, these lesions may be repaired adequately using pericardial patches. Pulmonary arteriovenous fistulae may develop in patients after a long-standing classic Glenn shunt. Rarely, large fistulae can be occluded with devices such as coils. More often, however, there are multiple small fistulae, which are not amenable to occlusion. Such lesions may regress after reparative surgery, but may be the source of severe, often lethal postoperative complications. In all patients, the pulmonary vascular bed (anatomy and physiology) must be evaluated very carefully, as abnormalities may preclude further surgery or be an important cause of failure after surgery in grown-ups with congenital heart disease.

### 5.1.6 Aortopulmonary collateral circulation

In cyanotic patients, aortopulmonary collateral vessels may complicate peri-operative and post-operative management. Important collateral circulation, if not curtailed, results in excessive return

to the left atrium. It obscures visualization of the operative field, washes out cardioplegia, compromises systemic perfusion and causes volume overload of the left side of the heart during the critical, post-operative period. The size, location and end-parenchymal distribution of the collateral vessels must therefore be established precisely. Adequate management may include (1) pre-operative occlusion (by interventional catheterization) or surgical ligation at the beginning of operation, (2) surgical unifocalization of large collateral vessels which are the sole sources of blood flow to significant portions of the lungs, (3) deep hypothermic low flow cardiopulmonary bypass to prevent the deleterious effects of multiple small collateral vessels not amenable to surgical occlusion.

## 5.2 Anaesthesia and post operative care

Grown-ups with congenital heart disease requiring cardiac and non-cardiac surgery present a spectrum of severity ranging from well patients with minor problems to those with extreme deviations from normal cardiovascular physiology. There is however, little evidence-based information to enable choice of anaesthetic technique in these patients. Anaesthesia demands particular attention to endocarditis prophylaxis, avoidance of air emboli, appropriate placement of vascular catheters and regulation of intra-vascular volume and systemic/pulmonary flow. The most important aspect of peri-operative care is the involvement of a clinical team with detailed understanding of the patient's cardiac defect, functional status and anticipated peri-operative stresses.

### 5.2.1 Physiology

Cardiovascular impairment and increased anaesthetic risk may be due to hypoxaemia, pulmonary vascular disease, cardiac failure or arrhythmia. Polycythaemia is the major adaptive response to hypoxaemia. Blood viscosity increases exponentially with haematocrit and may be further increased with iron deficiency.<sup>59</sup> Polythaemic patients must not become dehydrated and should receive intravenous fluids from the night before surgery. In patients with decreased pulmonary blood flow, hypoxaemia should be minimized by ensuring adequate hydration, maintaining systemic arterial pressure, minimizing transient elevation in pulmonary vascular resistance and avoiding increases in oxygen consumption. Elevated pulmonary blood flow, on the other hand, may increase

excessively cardiac work or decrease systemic perfusion. The strategy in this situation is to maintain ventricular performance and optimise pulmonary to systemic flow ratio. In the presence of a systemic to pulmonary shunt (e.g. Blalock-Taussig) pulmonary flow will vary with the pressure gradient. Increases in pulmonary blood flow decrease pulmonary compliance and increase airway resistance and work of breathing. Significantly increased pulmonary blood flow leads to airway obstruction and non-compliant lungs. Reactive pulmonary vasculature can be treated with anaesthetic drugs, positive pressure hyperventilation, oxygen and pulmonary vasodilators. If pulmonary blood flow is reduced, it is important to prevent further reductions. Airway dead space volume is increased with positive pressure ventilation, increased alveolar pressure or decreased left atrial or pulmonary artery pressures.

### 5.2.2 Assessment

Pre-operative assessment should define baseline problems and identify patients at increased risk. The cardiac anaesthetist should be involved in pre-operative conferences and overall planning of the management strategy. Access to all previous operation notes is invaluable. Clinical evaluation should be supplemented by laboratory data, ECG, chest X-ray, echocardiography and catheterization information. Lung function tests should be performed if the patient has scoliosis. Previous surgery may have resulted in recurrent laryngeal and phrenic nerve injuries or Horner's syndrome.

### 5.2.3 Anaesthetic management

Sedative pre-medication is popular. Oxygen consumption is reduced although caution must be exercised in the presence of hypoxaemia. Selection of induction agent is an individual choice. While there is a well-known influence of cardiac shunts on rapidity of inhalation induction, the clinical significance is rarely large. Vasodilatation is common during most induction techniques and will increase right to left shunting. However, the effect of this on arterial saturation will be partially offset by the reduction in total oxygen consumption and consequent increased mixed venous oxygen saturation. In general, the choice of drugs is less important than appropriate haemodynamic goals. Narcotic based anaesthesia is quite suitable in the presence of ventricular dysfunction. Ketamine may depress cardiac function if maximal sympathetic stimulation is present.

**Table 2** Energy expenditure, dynamic and static components in sports

Examples of sports	Energy expenditure in kcal/h (for 70 kg body weight)	static <sup>a</sup>	dynamic <sup>a</sup>
Light (<3 Met)			
Slow walking	200	-	+
Hiking	400	-	+
Golf	350	-	-
Cricket	350	-	+
Curling	300	-	-
Brisk walking	350	-	+
Gymnastics	400	++	-
Moderate (3–6 Met)			
Tour cycling	400	+	++
Swimming	500	+	++
Walking uphill with load	500	-	+
Horseback riding	200/450/550	+	-
walk/trot/gallop		+	-
Diving	450	++	+
Alpine skiing	350	-	+
Volleyball	350	-	+
Tennis double	400		
Hard (>6 Met)			
Tennis single	600	-	++
Jogging	700	+	+
Cross country skiing	700	-	++
(classic)	800	+	++
Cross country skiing	800	++	++
(skating)	800	++	++
Race cycling	500	+	+
Rowing	600	+	++
Boxing	700	++	++
Team handball, football			
Race Skating			

<sup>a</sup> - low, + moderate, ++ high.

#### 5.2.4 Monitoring

The use of invasive monitoring depends on the magnitude of surgery and the underlying cardiovascular pathophysiology. Placement of a pulmonary artery catheter can be technically difficult on account of anatomical abnormalities and might actually be dangerous in the presence of reactive pulmonary vasculature. Transoesophageal echocardiography is useful for following ventricular performance, valve function and blood flow. Some practical considerations for monitoring are important. End tidal CO<sub>2</sub> will underestimate paCO<sub>2</sub> in the presence of right to left shunting or common mixing. Previous systemic to pulmonary shunt placement requires blood pressure monitoring on the contra-lateral side. Accuracy of pulse oximeters is not guaranteed below levels of 80%. Vascular access may be problematic due to venous thrombosis or interrupted vena cava with azygos continuation. Central venous necklines pose a significant thromboembolic risk in the Fontan circulation and should

be removed as soon as feasible. Femoral veins can be used for central drug administration.

#### 5.2.5 Post anaesthesia care

Post operative care is usually provided in a high dependency or intensive care environment and standard management principles apply to most patients. Experience in the management of pulmonary vascular resistance, shunt lesions, ventricular outflow tract obstruction and right ventricular dysfunction is however particularly important. There is a trend in modern intensive care units to emphasise fluid administration and systemic oxygen delivery in high-risk surgical patients. This approach may not be well-tolerated by GUCH patients with poor ventricular function.

Cardiopulmonary interaction is important. Control of arterial blood gases (particularly PaCO<sub>2</sub>) is fundamental to regulating pulmonary blood flow. While low airway pressures and weaning from ventilation are beneficial to trans-pulmonary flow, it is

important to remember that the duration of the inspiratory phase during positive pressure ventilation has more influence than the absolute level of peak inspiratory pressure. Indeed shortening the inspiratory time (with consequent increase in peak inspiratory pressure) is usually the best strategy to maximise pulmonary flow. The alveolar ventilation  $\text{CO}_2$  response curve is normal. Hence adequate analgesia is important and appropriate. This is particularly important in the presence of labile pulmonary artery pressures. The hyperbolic relationship between  $\text{SaO}_2$  and  $\text{Qp:Qs}$  is often not understood by general intensive care staff. As a result, the significance of inappropriately high saturations in a complete mixing circulation may not be appreciated or raise concern.

### 5.3 Non-cardiac surgery

The risks of noncardiac surgery depend on the nature of the underlying cardiovascular abnormality, the extent of the surgical procedure, and whether or not it is an elective procedure or an urgent one. Preoperative planning must include consultation with a specialist GUCH centre.<sup>60</sup> The precise haemodynamic and anatomic abnormalities need to be understood, and the evaluation usually includes a Doppler echo assessment of ventricular function and pulmonary artery pressure. The risks of the operation must be explained to the surgeon including the risks of haemodynamic instability, haemorrhage, hypotension, and hypovolaemia, along with the risk of endocarditis.<sup>61</sup> Additional coexistent problems, which frequently accompany congenital heart disease, such as renal dysfunction, must also be evaluated in addition to those acquired disorders to which adult patients are vulnerable (systemic hypertension, ischaemic heart disease, arrhythmia (both ventricular and supraventricular), peripheral varicosities, etc.). Planning the procedure carefully is of the utmost importance. Urgent noncardiac operations carry a higher risk, and complications may be more frequent in patients undergoing respiratory or nervous system procedures. Intraoperative monitoring may be helpful in detecting early haemodynamic changes so that appropriate treatment can be initiated promptly. Continuous intra-arterial monitoring enables sudden changes in intravascular volume and haemodynamics to be detected, and facilitates periodic arterial blood gas determinations. The decision of whether or not to utilize a central venous line or pulmonary artery pressure recording (with or without oximetry) must be determined in each individual case and the risk-to-benefit ratio assessed. The latter may be associated with an

increased risk of ventricular arrhythmia and paradoxical embolism in patients with pulmonary hypertension and right-to-left shunt.

#### 5.3.1 Unoperated congenital heart disease

Patients with significant obstructive lesions (aortic stenosis, coarctation, and pulmonary stenosis) are vulnerable to intraoperative hypotension especially if ventricular function is depressed. Rapid fluid infusion may precipitate pulmonary edema. Elective noncardiac surgery may be performed more safely after repair or replacement of the obstructive valve or arterial lesion. A patient with ventricular dysfunction from any cause is more vulnerable to perioperative complications (e.g., congenitally corrected transposition of the great arteries when the systemic ventricle is the morphologic right ventricle).

#### 5.3.2 Operated congenital heart disease

Patients with repaired congenital heart disease are vulnerable to arrhythmia and haemodynamic deterioration, particularly when they have impaired ventricular function, which may be exacerbated by the loss of atrial transport. A multidisciplinary approach to their perioperative management will help to minimize the complications.

#### 5.3.3 Cyanotic heart disease

Patients with cyanotic congenital heart disease are most at risk from noncardiac surgery, especially if they have pulmonary hypertension.<sup>60</sup> The increased risk of haemorrhage secondary to the inherent haemostatic abnormalities can be temporarily reduced by preoperative phlebotomy, if the haematocrit is  $>65\%$ . One unit accompanied by isovolumic fluid replacement may be performed, and the blood can be saved for potential autologous transfusion. For cyanotic patients a fall in systemic vascular resistance can increase the right-to-left shunt and increase hypoxia. This is not tolerated well and can potentiate cardiovascular collapse. Lengthy operations associated with haemodynamic instability necessitating large volume fluid replacements are also associated with increased perioperative mortality. Spinal anaesthesia, similarly, can result in a fall in systemic vascular resistance and reduced venous return. This method of anaesthesia is thus best avoided in cyanotic patients. For any patient with a right-to-left shunt, there is an increased risk of a paradoxical embolus and air filters should be placed on all intravenous lines. Bacterial endocarditis prophylaxis should be given when appropriate.<sup>60,61</sup>

## 5.4 Transplantation (Fig. 5)

Transplantation is the final palliation for many grown-up patients with congenital heart disease and must be considered when the short-term prognosis is reduced or the quality of life unacceptable. Transplantation may involve either heart, heart-lung, single or double lung with intracardiac repair.

The most common congenital lesions that ultimately may require transplantation include failed Fontan, Mustard or Senning procedures, congenitally corrected transposition, complex pulmonary atresia and Eisenmenger complex. In addition, an increasing number of recipients will include those requiring retransplantation after primary transplantation for congenital lesions in childhood.

Risk stratification scores are available for terminal congestive heart failure<sup>62</sup> but may not apply to grown-ups with congenital heart disease. This makes decisions regarding timing of transplantation difficult.

In addition to standard pretransplant work up, attention needs to be directed towards specific issues in grown-ups with congenital heart disease. Sensitization from previous transfusions may increase the early risk of rejection and graft failure. Treatment to reduce the HLA antibody level may be required before placement on the waiting list. Assessment of pulmonary vascular resistance is difficult in patients with low cardiac output, residual lesions, shunts or collaterals. Detailed planning of the surgical approach is crucial in many complex lesions and all anatomic details, including the systemic and pulmonary venous return need to be defined. MRI is particularly informative. Non-adherence to post transplant medications is a major problem in young adults and pre-existing psychosocial issues may require careful evaluation before listing. Other risk factors that are specific to the failing 'Fontan' patient include protein losing enteropathy and pulmonary arteriovenous fistulae. An increasing number of patients with terminal cardiopulmonary lesions have underlying chromosomal anomalies, which may further complicate decision-making.

Surgical issues in previously operated patients include difficulties with cannulation, dissection, abnormal anatomy, need for an additional conduit for reconstruction, bleeding and prediction of the time needed to prepare the recipient. This often leads to longer ischaemia and bypass times, which may jeopardize graft function and survival.

The current 1-, 5- and 10-year survival after cardiac transplantation in non-GUCH patients is 80, 70 and 55%.<sup>29</sup> For heart-lung transplantation the

survival is only 65, 45 and 30%, respectively.<sup>29</sup> The outcome after heart transplantation is significantly lower in many grown-ups with congenital heart disease, including Fontan patients, primarily due to a higher early attrition.<sup>63–66</sup> Late attrition after transplantation has not improved significantly during the last 10–20 years despite introduction of new immunosuppressants. Chronic rejection (graft coronary vasculopathy in hearts and obliterative bronchiolitis in lungs) and malignancy remain the main concerns. Immunologic progress and new drugs will hopefully improve this in the future.

The increasing donor shortage and inferior results may influence donor organ allocation to grown-ups with congenital heart disease patients. The increasing number of patients surviving with palliated lesions and those with previous transplants will worsen the donor situation dramatically and many patients will never get to transplantation until viable alternatives such as reliable long-term mechanical support systems or xenotransplantation become available. This problem is of particular concern for grown-ups with congenital heart disease requiring heart-lung grafts.

## 6 Psychosocial issues (Fig. 2)

In addition to provision of care for complex medical and surgical problems, the specialist service for grown-ups with congenital heart disease must provide support for the many psychosocial problems in this population.<sup>67</sup> These include a high level of anxiety about the underlying heart condition and prognosis, difficulties with social interaction as well as specific issues regarding employment, insurance and physical activity.<sup>68</sup> Staff with appropriate expertise should form part of the specialist team and are frequently called upon to act as advocates for the patients who may face unfair discrimination. Further studies are required to investigate the relationship between the underlying disability, level and type of follow up care, emotional status and affect on psychosocial health and performance. These should examine well-characterized populations of patients, use validated questionnaires, grading of physiological performance and appropriate control groups for comparison. Currently, validated congenital heart disease specific measures are not available. This information will assist in the training of professionals and enable the provision of counselling services, specific to the needs of adolescents and adults with congenital heart disease.

## 6.1 Intellectual development/education

Intellectual development may be influenced by genotype, the presence of syndromes, as well as the disturbed haemodynamics of the cardiac defect and its treatment. Many patients with syndromes involving learning disabilities of varying degree now survive into adult life, including William's syndrome, Down's syndrome and 22q11 deletion. They have specific educational needs. Cognitive function may be affected by early neurological complications of low cardiac output, acidosis, and hypoxia as well as by the consequences of cardiac surgery, anaesthesia, hypothermia, circulatory arrest and cardiopulmonary bypass. Intellect may be further influenced by chronic cardiac failure, arrhythmia and/or cyanosis as well as absence from school.

Studies of intellectual outcome in young adults with relatively simple congenital heart defects have been very encouraging. Indeed, level of educational attainment may be superior to that in the general population, reflecting the high level of motivation of grown-ups with congenital heart disease and the support of their families and health care providers.<sup>69</sup> Data in patients with complex defects are more limited. Some studies have shown below average I.Q. scores in cyanotic congenital heart disease, including Tetralogy of Fallot and transposition of the great arteries.<sup>70</sup> Individual factors such as duration of chronic hypoxia and elevated haematocrit may have a limited impact but, in general, combinations of adverse factors appear to have long-term detrimental effects. Up to 10% of children with congenital heart disease will have a cerebrovascular infarction, most within the first 2 days of life.<sup>71</sup> Early correction of congenital heart disease, as is currently undertaken, may reduce these neurological complications, but may in itself, have neurocognitive effects which will become manifest in later life.

## 6.2 Employment

Ability to obtain and maintain employment will depend on intellectual and physical capacity, motivation, and interaction with peers as well as potential discrimination by society. Several reports of employment status of grown-ups with congenital heart disease have shown that unemployment is more common in patients with complex lesions.<sup>72</sup> Approximately 10% are considered totally disabled. The economic consequences of unemployment vary in different countries, because of the marked differences in the levels of social and welfare programmes across Europe. Nevertheless, even under the most generous systems, unemployment has

major adverse effects including lowered self-esteem and social contact, especially in this potentially vulnerable population.

Appropriate employment counselling is part of the responsibility of the medical profession, assisted by other members of the specialist team, including nurse counsellors. Advice needs to be realistic and based on physical and intellectual capacity as well as on specific issues, such as arrhythmia, which may preclude certain types of job (e.g. car driving). There are often specific exclusions for jobs, particularly in the public sector, which varying between countries, (e.g. armed forces and police). The ability of the patient to undertake the demands of different occupations may need to be tested in formal programmes. Other services such as vocational and physical rehabilitation as well as job training may broaden the patient's range of employment options.

Studies have shown discrimination against job applicants with congenital heart disease. These are based on concerns about performance, absenteeism, premature retirement and medical insurance. Legislation against such behaviour by potential employers varies between countries, including affirmative action to employ disabled persons. Furthermore, prohibition of discrimination on the basis of higher insurance costs is often included and is crucial in countries with an insurance-based system for medical care. The practitioner can greatly increase patients' employment chances by interacting directly with the prospective employer, as objective outcome data for many conditions are still not available.

## 6.3 Insurance

Life insurance and health insurance availability vary greatly both within and between countries. Despite attempts to standardize and improve the situation, patients may still need to shop around to obtain the best deal. Physicians and organized patient groups, such as the Grown-Up Congenital Heart Association in the UK, provide extremely useful advice and assistance in directing patients to 'friendly' companies.

Life insurance remains an important component of financial planning, not least in relation to mortgage and house purchase. Life insurance may be denied, be made available at normal rates or have a heavily loaded additional premium.<sup>73</sup> A study in 1993 in the UK evaluated both life and health insurance available to adults with congenital heart disease.<sup>74</sup> The findings revealed a surprising discordance between insurance policy and medical

outcome data. For example, patients with repaired aortic coarctation were insurable at normal rates, whilst patients were often denied insurance after successful repair of ventricular septal defect. Health insurance was usually available, but almost always excluded benefit for the underlying cardiac condition. This can have a major impact on delivery of optimal medical care to grown-ups with congenital heart disease in an insurance-based medical system. In an earlier German survey among a cohort of GUCH patients, more than 30% were refused life insurance.<sup>75</sup> Further studies are in progress in the UK and France to obtain up to date information. Insurance recommendations from cardiac societies exist in Italy and Switzerland, but not in the other European countries surveyed. This is clearly an unsatisfactory situation and could be improved by establishment of formal practice guidelines and outcome analysis, which could be shared with insurers. Strategies for insurance of patients with more complex lesions and worse outcomes need to be developed if medical care is not to be compromised by financial considerations.

#### 6.4 Physical activity/sport

Participation in sports and regular physical exercise have well documented beneficial effects on fitness, psychological well-being, confidence and social interaction as well as on the later risk of acquired cardiac disease. Recommendations on exercise in grown-ups with congenital heart disease need to be based on the ability of the patient as well as on the impact of physical training on cardiac haemodynamics<sup>76,77</sup> (e.g. ventricular remodelling, myocardial ischaemia). Counselling should include an appreciation of the type of energy expenditure involved in different sports and teaching of a method to enable the patient to limit his or her activities (Table 2). These include the Borg scale of perceived effort, a target heart rate range (60–80% of maximum heart rate achieved during testing without symptoms or haemodynamic deterioration) and a simple breathing rule (activity can be carried out safely as long as breathing still permits comfortable speech). Impact sport should be avoided in patients with Marfan's syndrome or other aortic anomalies, those on oral anticoagulants or those with pacemakers. Formal testing, assessing the impact of exercise levels relevant to the patients' expectations during normal day life, should be undertaken and protocols derived from conventional adult exercise testing programmes need to be adapted.<sup>78</sup> Doctors should use these results in discussion with patients. In general, medical practitioners are conservative and are often

unnecessarily proscriptive in their recommendations; this may have important adverse effects on quality of life.

Exercise may have acute, chronic and potentially harmful haemodynamic effects in patients with congenital heart disease. These include fluid depletion, blood pressure rise or fall, tachycardia and/or arrhythmia as well as long-term effects on ventricular hypertrophy and function. Of most concern is the risk of sudden death during or after exercise. Most cases of sudden death during physical activity in the young are due to a previously unrecognized cardiac disorder and sudden death in patients with known congenital heart disease is very rare (1 in 10 000 patients in a recent survey in nine specialist centres for grown-ups with congenital heart disease). Potentially lethal situations may occur with arrhythmia and haemodynamically vulnerable circulations (e.g. preload jeopardized circuits such as after Mustard/Senning or Fontan operation or with heart failure. Advice to perform social exercise to a level of comfort, but not to attempt competitive sports is applicable in most situations.<sup>79,80</sup>

#### 6.5 Quality of life

Few studies of quality of life have been performed using validated measures in large populations of adolescent or adults with congenital heart disease. Most have limited relevance to modern management.<sup>81</sup> Most patients, when questioned, will say that they are 'asymptomatic'. Whilst many are able to enjoy the full range of normal life activities, patient symptom reporting should be interpreted with caution as they may always have been limited and thus 'do not know any better'.<sup>82</sup> Alternatively, they may be reluctant to admit limitations and to discuss problems. Allocation of sufficient time to obtain a good history (often by the nurse counsellor) is essential and may need to be supplemented by objective testing.

Psychosocial adjustment to adult life depends, not only on the type and severity of the congenital heart defect, but also on the attitude and behaviour of family, friends and the GUCH team. Sensitive handling and education can be enormously valuable and the advocacy role of the GUCH team cannot be over emphasized.

#### 6.6 Patient organizations

In many European countries, patient organizations have been established and are active in spreading medical information, educating patients and securing patient rights. These organizations, due to their

contact with media, may often have a greater political impact than normal health channels. Patient specific websites are invaluable for education as well as helping to direct patients to appropriate specialist centres. Many patient

organizations have been effective in raising money for medical research and new expensive treatment approaches. Regular contact between specialist units for grown-ups with congenital heart disease and these organizations is therefore important.

## 7 Specific lesions

This section summarises the current management strategy for the commonest lesions seen in grown-ups with congenital heart disease. Many recommendations are based at clinical experiences rather than evidence trial randomized clinical trials. We have therefore chosen not to use categories of strength of endorsement as in other ESC Task Forces.

### Atrial septal defect

Criteria	Comments	
1. Introduction and background	● common defect which may be diagnosed first in adult life	
2. Survival → adult life	● small defects-excellent prognosis ● large defects – reduced survival, depending on age at treatment	
3. Haemodynamic issues	● PHT ● RV dilation/failure ● potential for paradoxical embolism ● reduced LV compliance	
4. Arrhythmia/pacing	● atrial arrhythmia (atrial fibrillation and flutter) ● sick sinus syndrome ● pacing rarely required	
5. Investigations	ECG	● baseline – if clinically indicated (arrhythmias)
	Chest X-ray	● baseline – otherwise little value
	ECHO/TOE	● baseline-location, size, RV size, PA pressure, Qp:Qs, associated lesions ● TOE usually performed in older patients and at device closure
	Catheterization	● device closure ● PVR assessment
	MRI	● rarely helpful
	Holter	● if symptomatic arrhythmia
	Exercise function	● baseline – little value
6. Indications for intervention	● large defects (>10 mm) unless pulmonary vascular disease (PVR>8 Um <sup>2</sup> , L-R shunt <1.5, no response to pulmonary vasodilators) ● paradoxical embolism	
7. Interventional options	● surgery or device closure (stretched diameter <38 mm)	
8. Post treatment outcome	● low risk procedure unless PVD ● late intervention less successful	
9. Endocarditis	● very rare ● prophylaxis not indicated	
10. Pregnancy/contraception/recurrence/fetal	● no contra-indications unless PVD ● no restrictions for contraception ● consider fetal echocardiography	
11. Recurrence/genetics	● 3% of first degree relatives ● familial ASD (with long PR interval) ● autosomal dominant	
12. Syndromes	● Holt Oram – upper limb deformity ● autosomal dominant	
13. Sport/physical activity	● no restrictions unless moderate/severe PVD	

14. Insurance	● category 1 ● generally no problem if defect closed early
15. Follow-up interval	● early repair (<30 years) – no problems – discharge ● late repair – regular f/u
16. Follow-up care	● level 2
17. Unresolved issues	● surgery vs device closure ● when to close in PHT ● concomitant Maze procedure ● upper age limit for surgery ● PFO closure in patients with suspected paradoxical embolism

### Ventricular septal defect – unrepaired

Criteria	Comments	
1. Introduction and background	● significant ventricular septal defects usually repaired in childhood ● see ventricular septal defect and PVD (Eisenmenger) but diminishing ● small ventricular septal defect or postoperative septal defect common in adults ● Eisenmenger patients becoming less frequent	
2. Survival → adult life	● excellent for small ventricular septal defect ● large ventricular septal defect may have pulmonary vascular disease (Eisenmenger) ● may develop aortic regurgitation	
3. Haemodynamic issues	● left-right shunt ● LV dilatation and impaired function ● aortic regurgitation ● pulmonary vascular resistance in uncorrected large ventricular septal defect	
4. Arrhythmia/pacing	● rare	
5. Investigations	Chest X-ray	● baseline-cardiomegaly
	ECG	● routine ● rhythm chamber enlargement
	ECHO	● number size and location of defects ● LV/RV function ● aortic regurgitation
	TOE	● if TTE image inadequate
	Catheter	● pulmonary vascular resistance ● associated lesions.
	MRI	● rarely helpful
	Holter	● only if symptomatic
	Exercise test	● only if symptomatic ● sports counselling
6. Indications for intervention	● left – right shunt with left heart volume overload ● reversible pulmonary hypertension ● aortic regurgitation ● associated abnormalities (RV outflow tract, subaortic stenosis) ● previous endocarditis	
7. Interventional options	● surgery ● catheter closure in muscular VSD(s)	
8. Post treatment outcome	● good surgical results	
9. Endocarditis	● prophylaxis in all	

10. Pregnancy/contraception	<ul style="list-style-type: none"> <li>● no contra-indications in uncomplicated VSD</li> <li>● pregnancy contra-indicated in pulmonary vascular disease (Eisenmenger disease)</li> </ul>
11. Recurrence/genetics	<ul style="list-style-type: none"> <li>● occasionally familial</li> <li>● usual recurrence risk</li> <li>● common cardiac anomaly in syndromes e.g. Down's</li> </ul>
12. Sport/physical activity	<ul style="list-style-type: none"> <li>● no restriction in small ventricular septal defect</li> </ul>
13. Insurance	<ul style="list-style-type: none"> <li>● small ventricular septal defects category 1</li> </ul>
14. Follow-up interval	<ul style="list-style-type: none"> <li>● infrequent follow-up unless haemodynamic abnormalities (e.g. aortic regurgitation)</li> </ul>
15. Follow-up care	<ul style="list-style-type: none"> <li>● small ventricular septal defect 3, pulmonary vascular disease (Eisenmenger) 2, aortic regurgitation/complicated haemodynamics 1</li> </ul>
16. Unresolved issues	<ul style="list-style-type: none"> <li>● optimal management of Eisenmenger patients</li> </ul>

### Repaired ventricular septal defect

Criteria	Comments	
1. Introduction and background	<ul style="list-style-type: none"> <li>● common lesion</li> <li>● most patients now adults</li> </ul>	
2. Survival – adult life	<ul style="list-style-type: none"> <li>● excellent survival</li> <li>● occasional residual shunt</li> <li>● some develop RV or LV outflow tract obstruction</li> <li>● some develop aortic regurgitation</li> </ul>	
3. Haemodynamic issues	<ul style="list-style-type: none"> <li>● residual shunt</li> <li>● ventricular function</li> <li>● aortic regurgitation</li> <li>● new haemodynamic abnormalities (RV outflow obstruction)</li> </ul>	
4. Arrhythmia/pacing	<ul style="list-style-type: none"> <li>● rare AV block, ventricular arrhythmia</li> </ul>	
5. Investigations	Chest X-ray	<ul style="list-style-type: none"> <li>● baseline – cardiomegaly</li> </ul>
	ECG	<ul style="list-style-type: none"> <li>● rhythm</li> </ul>
	Echo	<ul style="list-style-type: none"> <li>● residual VSD(s)</li> <li>● LV/RV function</li> <li>● aortic regurgitation</li> </ul>
	TOE if TTE insufficient	<ul style="list-style-type: none"> <li>● TOE only if TTE inadequate</li> </ul>
	Catheter	<ul style="list-style-type: none"> <li>● rarely required</li> </ul>
	MRI	<ul style="list-style-type: none"> <li>● rarely helpful</li> </ul>
	Holter	<ul style="list-style-type: none"> <li>● only if symptomatic</li> </ul>
	Stress test	<ul style="list-style-type: none"> <li>● only if symptomatic</li> <li>● sports counselling</li> </ul>
6. Indications for intervention	<ul style="list-style-type: none"> <li>● if residual VSD; see 'unrepaired VSD'</li> </ul>	
7. Interventional options	<ul style="list-style-type: none"> <li>● see 'unrepaired VSD'</li> </ul>	
8. Post treatment outcome	<ul style="list-style-type: none"> <li>● see 'unrepaired VSD'</li> </ul>	
9. Endocarditis	<ul style="list-style-type: none"> <li>● prophylaxis if residual VSD</li> <li>● questionable in closed VSD</li> </ul>	
10. Pregnancy/contraception	<ul style="list-style-type: none"> <li>● no contra-indications in uncomplicated closed VSD</li> <li>● Pregnancy contra-indicated in PVOD (Eisenmenger)</li> </ul>	
11. Recurrence/genetics	<ul style="list-style-type: none"> <li>● see: 'unrepaired VSD'</li> </ul>	

12. Sport/physical activity	● no restriction in closed VSD
13. Insurance	● category 1
14. Follow-up interval	● can discharge if closed VSD without any residual abnormalities ● infrequent follow-up for minor residual lesions
15. Follow-up care	● Eisenmenger 2, small VSD 3, aortic regurgitation/complicated haemodynamic 1
16. Unresolved issues	—

#### Postoperative complete atrio-ventricular septal defect

Criteria	Comments	
1. Introduction and background		
2. Survival → adult life	● unoperated survivors develop PVD ● surgical results markedly improved ● status after repair depends mostly on left AV valve function ● many patients have Down's syndrome	
3. Hemodynamic issues	● left AV-valve regurgitation (±stenosis) ● pulmonary vascular disease ● late sub-aortic stenosis	
4. Arrhythmia/pacing	● risk of complete heart block low (<2%) ● atrial arrhythmias especially with left AV-valve dysfunction	
5. Investigations	Chest X-ray	● cardiomegaly ● pulmonary vascular markings ● pulmonary vascular disease
	ECG	● routine (LVH, RVH, CVH) ● superior QRS-axis ● right bundle branch block ● conductance disturbances
	ECHO/TOE	● most useful investigation for ● left AV valve morphology and function ● ventricular function ● residual lesions (shunt, sub-aortic stenosis)
	Catheter	● rarely required unless reoperation considered
	MRI	● rarely indicated
	Holter	● only in symptomatic patients
	Exercise testing	● rarely indicated
	Additional investigations	● significant left AV valve dysfunction ● significant residual shunt ● subaortic stenosis
6. Indications for reintervention	● significant left AV-valve dysfunction ● significant residual shunt ● sub-aortic stenosis ● progressive/symptomatic AV-Block	
7. Interventional options	● re-operation may require valve replacement	
8. Post treatment outcome	● excellent long-term results unless ● actuarial survival after 20 years >80% ● left AV valve regurgitation (stenosis) ● pulmonary vascular disease ● late sub-aortic stenosis	
9. Endocarditis	● prophylaxis in all cases	

10. Pregnancy/contraception	<ul style="list-style-type: none"> <li>● pregnancy contra-indicated in PVD (Eisenmenger)</li> <li>● anticoagulation management in patients with prosthetic valves</li> <li>● avoid oestrogen containing pill in pulmonary hypertension</li> </ul>
11. Recurrence/genetics/syndromes	<ul style="list-style-type: none"> <li>● above average recurrence risk Down's syndrome in &gt;50% of complete AVSD</li> <li>● app. 10–14% CCD in mothers with AVSD</li> </ul>
12. Physical activity/sports	<ul style="list-style-type: none"> <li>● no restrictions if good repair and no significant arrhythmias</li> </ul>
13. Insurance	<ul style="list-style-type: none"> <li>● category 2 if well repaired</li> </ul>
14. Follow-up interval	<ul style="list-style-type: none"> <li>● 1–2 yearly intervals with ECG and ECHO in stable cases</li> </ul>
15. Follow-up care	<ul style="list-style-type: none"> <li>● level 2 unless significant haemodynamic problems</li> </ul>
16. Unresolved issues	<ul style="list-style-type: none"> <li>● only limited data regarding long term prognosis</li> </ul>

### Postoperative partial atrio-ventricular septal defect (p-AVSD)

Criteria	Comments	
1. Introduction and background		
2. Survival → adult life	<ul style="list-style-type: none"> <li>● similar to secundum atrial septal defect unless significant to left AV-valve regurgitation</li> <li>● unoperated p-AVSD have reduced life expectancy</li> <li>● PVOD may develop late</li> <li>● status after repair depends on left AV valve function</li> </ul>	
3. Hemodynamic issues	<ul style="list-style-type: none"> <li>● before repair</li> <li>● size of shunt</li> <li>● degree of AV valve regurgitation</li> <li>● after repair</li> <li>● residual shunt and left AV valve regurgitation</li> <li>● subaortic stenosis</li> </ul>	
4. Arrhythmia/pacing	<ul style="list-style-type: none"> <li>● atrial arrhythmias rare unless left AV-valve regurgitation</li> <li>● complete heart block very rare</li> <li>● pacing rarely required unless sick sinus syndrome</li> </ul>	
5. Investigations	ECG	<ul style="list-style-type: none"> <li>● superior-QRS axis</li> <li>● right bundle branch block</li> <li>● rhythm follow-up</li> </ul>
	Chest X-ray	<ul style="list-style-type: none"> <li>● routine</li> <li>● cardiomegaly</li> </ul>
	ECHO/TOE	<ul style="list-style-type: none"> <li>● most useful investigation both for and after operation</li> <li>● left AV valve function</li> <li>● ventricular function</li> <li>● residual lesion</li> </ul>
	Catheter	<ul style="list-style-type: none"> <li>● rarely required unless reoperation considered</li> </ul>
	MRI	<ul style="list-style-type: none"> <li>● rarely indicated</li> </ul>
	Holter	<ul style="list-style-type: none"> <li>● rarely indicated</li> </ul>
	Exercise testing	<ul style="list-style-type: none"> <li>● rarely indicated</li> </ul>
	Additional investigations	—
6. Indications for intervention	<ul style="list-style-type: none"> <li>● all cases to be considered for intervention unless pulmonary vascular disease</li> <li>● reoperation for significant left AV-valve regurgitation</li> <li>● residual shunt or subaortic stenosis</li> <li>● progressive/symptomatic arrhythmias</li> </ul>	

7. Interventional options	<ul style="list-style-type: none"> <li>● surgery with valve repair or replacement</li> <li>● closure of re-/residual ASD</li> <li>● Pacemaker (DDD) in progressive/symptomatic complete block</li> </ul>
8. Outcome	<ul style="list-style-type: none"> <li>● excellent long term provided left AV-valve repair satisfactory</li> </ul>
9. Endocarditis	<ul style="list-style-type: none"> <li>● prophylaxis indicated if left AV-valve regurgitation is present</li> </ul>
10. Pregnancy/contraception	<ul style="list-style-type: none"> <li>● well tolerated in repaired cases</li> <li>● contraindicated in rare cases with PVOD (Eisenmenger)</li> <li>● anticoagulation management in patients with prosthetic valves</li> <li>● avoid oestrogen containing pill in pulmonary hypertension</li> </ul>
11. Recurrence/genetics/syndromes	<ul style="list-style-type: none"> <li>● none</li> </ul>
12. Physical activity/sports	<ul style="list-style-type: none"> <li>● no restrictions if good repair and no significant arrhythmias</li> </ul>
13. Insurance	<ul style="list-style-type: none"> <li>● category 2</li> </ul>
14. Follow-up interval	<ul style="list-style-type: none"> <li>● 2 yearly intervals with ECG and ECHO in stable cases</li> </ul>
15. Follow-up care	<ul style="list-style-type: none"> <li>● unoperated level 1, postoperative level 2</li> </ul>
16. Unresolved issues	<ul style="list-style-type: none"> <li>● long term function of a non-reconstructed AV-valve is uncertain</li> </ul>

### Pulmonary stenosis

Criteria	Comments	
1. Introduction and background		
2. Survival → adult life	<ul style="list-style-type: none"> <li>● excellent if relieved effectively</li> <li>● poor if severe valve PS untreated</li> </ul>	
3. Haemodynamic issues	<ul style="list-style-type: none"> <li>● PS severity</li> <li>● PR severity</li> <li>● leaflet dysplasia</li> <li>● right ventricular function</li> </ul>	
4. Arrhythmia/pacing	<ul style="list-style-type: none"> <li>● atrial arrhythmias in RV failure and tricuspid regurgitation</li> <li>● pacing not indicated</li> </ul>	
5. Investigations	Chest X-ray	<ul style="list-style-type: none"> <li>● baseline otherwise little value unless RV failure</li> </ul>
	ECG	<ul style="list-style-type: none"> <li>● rhythm RV</li> <li>● hypertrophy</li> </ul>
	ECHO/TOE	<ul style="list-style-type: none"> <li>● investigation of choice for RVOT gradient</li> <li>● pulmonary regurgitation</li> <li>● RV size/function</li> <li>● tricuspid regurgitation</li> </ul>
	Catheter	<ul style="list-style-type: none"> <li>● rarely needed except for balloon dilatation</li> </ul>
	MRI	<ul style="list-style-type: none"> <li>● rarely needed</li> <li>● assess RV size/function and RA dilation in severe pulmonary regurgitation</li> </ul>
	Holter	<ul style="list-style-type: none"> <li>● not routinely indicated</li> </ul>
	Exercise	<ul style="list-style-type: none"> <li>● not routinely indicated</li> </ul>
	Additional investigations	<ul style="list-style-type: none"> <li>● none</li> </ul>
6. Indications for intervention	<ul style="list-style-type: none"> <li>● valve gradient &gt;30 mmHg at rest or for symptoms</li> </ul>	

7. Interventional options	● balloon valvuloplasty almost always ● surgery if valve calcified/dysplasty
8. Post treatment outcome	● excellent long-term results unless early failure significant pulmonary regurgitation uncommon
9. Endocarditis	● low risk. Prophylaxis may not be required in mild cases
10. Pregnancy/contraception/fetal	● routine pregnancy unless moderate to severe PS or right to left shunt through ASD or PFO
11. Recurrence/genetics	● 4% approximately
12. Syndromes	● Noonan ● congenital rubella ● Williams ● Alagille
13. Sport/physical activity	● unrestricted unless severe
14. Insurance	● category 1 after successful treatment or mild PS
15. Follow-up interval	● can discharge if mild with ECHO. Every 1–3 years if more than mild, PR, or desaturation.
16. Follow-up care	● mild PS: 3, Excellent early result: 2, Residual gradient or significant PR: 2
17. Unresolved issues	● none

## Tetralogy of fallot – postoperative

Criteria	Comments	
1. Introduction and background	● common lesion. Most Fallot patients are now adults	
2. Survival → adult life	● survival rate after surgery excellent (normal in selected groups) ● occasionally unoperated patients survive into adulthood.	
3. Haemodynamic issues	● pulmonary regurgitation/PS and RV function ● tricuspid regurgitation ● aortic regurgitation ● residual lesions	
4. Arrhythmia/pacing	● late complete heart block rare ● ventricular premature beats common in asymptomatic patients ● symptomatic VT rare ● atrial arrhythmias common and relate to poor haemodynamics ● small incidence of late sudden death	
5. Investigations	Chest X-ray	● baseline and occasionally follow-up ● cardiomegaly ● RV outflow
	ECG	● routine ● rhythm ● access/QRS width (usually complete right bundle branch block)
	ECHO/TOE	● regularly for PR/RVOTO/RV size function/tricuspid regurgitation? Aortic regurgitation/LV function
	Catheter	● preoperative for residual lesions, coronary anatomy intervention for dilatation/stent of pulmonary arteries ● possibly in future for implantable pulmonary valve
	MRI	● may become investigation of choice for RV size function and pulmonary regurgitation
	Holter	● for symptoms and in poor haemodynamics

	Exercise	● exercise capacity, arrhythmias
	Additional investigations	● electrophysiological study for syncope, sustained arrhythmia (atrial or ventricular), RFA
6. Indications for intervention		● significant RVOT or PA branch stenosis ● aortic regurgitation ● residual VSD, significant pulmonary regurgitation (with symptoms and RV dilatation)
7. Interventional options		● surgery, surgery with ablation, balloon dilatation/stenting, RF ablation catheter intervention for pulmonary valve insertion
8. Post treatment outcome		● most patients well ● RV function may not normalize after pulmonary valve replacement ● arrhythmia may persist ● risk of sudden death
9. Endocarditis		● prophylaxis in all
10. Pregnancy/contraception/fetal		● no contra-indication to pregnancy in well repaired patients ● monitor ventricular function and arrhythmia ● no additional fetal risk
11. Recurrence/genetics		● 1.5% for father, 2.5–4% for mother with TOF, 16% of Fallot patients have deletion of chromosome 22q11- recurrence risk 50%.
12. Syndromes		● 22q11
13. Sport/physical activity		● no contra-indication to sport unless documented arrhythmia ● significant ventricular dysfunction
14. Insurance		● category 2
15. Follow-up interval		● one/two yearly with ECG, ECHO±Holter, exercise test
16. Follow-up care		● 1 if documented residual abnormalities/arrhythmia, 2 otherwise
17. Unresolved issues		● risk stratification for sudden death ● indication for implantable defibrillator ● timing of reoperation for pulmonary regurgitation

### Conduits

Criteria	Comments	
1. Introduction and background	● conduits used in repair of complex congenital heart disease ● usually RV-PA (e.g. PA/VSD, Truncus, ToF, TGA/VSD/PS)	
2. Survival → adult life	● all conduits in children deteriorate and require replacement (usually <10 years) ● longevity of replacement unclear	
3. Haemodynamic issues	● stenosis of valve, subvalve or anastomosis to PA ● pulmonary regurgitation with RV volume overload ● LV-Aortic pathway in complex repairs	
4. Arrhythmia/pacing	● ventricular arrhythmias, surgical heart-block	
5. Investigations	Chest X-ray	● baseline and follow-up ● conduit calcification ● cardiomegaly
	ECG	● routine ● rhythm

	ECHO	<ul style="list-style-type: none"> <li>● investigation of choice for follow-up of RV</li> <li>● pressure gradient across conduit and PI</li> <li>● LV aortic pathway</li> <li>● ECHO may underestimate gradient</li> </ul>
	TOE	<ul style="list-style-type: none"> <li>● not routine</li> </ul>
	Catheterization	<ul style="list-style-type: none"> <li>● evaluation for surgery</li> <li>● balloon dilatation or stenting</li> </ul>
	MRI	<ul style="list-style-type: none"> <li>● very useful for investigation of conduit function</li> <li>● may become investigation of choice</li> </ul>
	Holter	<ul style="list-style-type: none"> <li>● only if arrhythmia suspected</li> </ul>
	Exercise function	<ul style="list-style-type: none"> <li>● not routine</li> <li>● useful for objective evaluation of exercise tolerance</li> </ul>
6. Indications for intervention	<ul style="list-style-type: none"> <li>● significant symptoms or conduit obstruction</li> </ul>	
7. Interventional options	<ul style="list-style-type: none"> <li>● usually surgical replacement of conduits</li> <li>● occasionally balloon dilatation or stenting</li> </ul>	
8. Post treatment outcome	<ul style="list-style-type: none"> <li>● fate of replaced conduit uncertain</li> <li>● need long term follow-up</li> </ul>	
9. Endocarditis	<ul style="list-style-type: none"> <li>● prophylaxis in all</li> </ul>	
10. Pregnancy/contraception	<ul style="list-style-type: none"> <li>● pregnancy tolerated if haemodynamics stable</li> <li>● no contraception issues</li> </ul>	
11. Recurrence/genetics	<ul style="list-style-type: none"> <li>● usual recurrence rate for congenital heart disease</li> <li>● higher if 22q11 deletion</li> </ul>	
12. Syndromes	<ul style="list-style-type: none"> <li>● 22q11 deletion</li> </ul>	
13. Sport/physical activity	<ul style="list-style-type: none"> <li>● avoid contact sports</li> <li>● otherwise no restrictions if haemodynamics good</li> </ul>	
14. Insurance	<ul style="list-style-type: none"> <li>● level 2</li> </ul>	
15. Follow-up interval	<ul style="list-style-type: none"> <li>● yearly with ECHO, ETT for ventricular function, arrhythmia surveillance (ECG, Holter if symptoms)</li> <li>● significant conduit dysfunction may be present in mildly symptomatic patients</li> </ul>	
16. Follow-up care	<ul style="list-style-type: none"> <li>● category 1</li> </ul>	
17. Unresolved issues	<ul style="list-style-type: none"> <li>● type of conduit (homograft versus xenograft)</li> <li>● role of balloon dilatation stenting</li> </ul>	

## Aortic valve stenosis (unoperated)

Criteria	Comments	
1. Introduction and background	<ul style="list-style-type: none"> <li>● common especially bicuspid aortic valve (1–2% of population)</li> <li>● may occur with other lesions</li> </ul>	
2. Survival → adult life	<ul style="list-style-type: none"> <li>● normal if mild obstruction</li> </ul>	
3. Hemodynamic issues	<ul style="list-style-type: none"> <li>● degree of stenosis may progress</li> <li>● associated aortic regurgitation</li> <li>● LV hypertrophy and function</li> </ul>	
4. Arrhythmia/pacing	<ul style="list-style-type: none"> <li>● VT and VF may occur during exertion with severe obstruction</li> </ul>	
5. Investigations	EKG	<ul style="list-style-type: none"> <li>● LVH and repolarization changes</li> </ul>

	Chest X-ray	● baseline ● calcification
	ECHO	● investigation of choice ● LV mass/function ● aortic valve/size/morphology/area ● LV to aortic gradient ● aortic regurgitation
	TOE	● rarely of value except in endocarditis
	MRI	● rarely of value
	Catheter	● not for diagnosis ● for coronary angiography and balloon dilatation
	Exercise testing	● for repolarization changes and symptoms ● surgical decision making
6. Indications for intervention	● symptoms: severe LV pressure overload ● severe aortic stenosis	
7. Interventional options	● balloon valvuloplasty if valve uncalcified ● rarely good option in adult ● mechanical valve replacement, homograft or Ross procedure depending on patient's age, sex, preferences and local expertise	
8. Outcome	● recurrence common late after valvotomy ● very good in uncomplicated cases of valve replacement.	
9. Endocarditis	Prophylaxis indicated in all	
10. Pregnancy/contraception	● low risk in asymptomatic patients even with moderate obstruction ● high risk in patients with severe obstruction ● transcatheter intervention may be indicated in unplanned pregnancy	
11. Recurrence/genetics/Syndromes	● bicuspid valve may be familial ● association with coarctation ● recurrence rate may be higher in syndromes.	
12. Physical activity/sports	● no competitive sports if obstruction is moderate or severe	
13. Insurance	● category 2	
14. Follow-up interval	● depends on severity and progression rate ECG/ECHO±exercise test	
15. Follow-up care	● mild 3-moderate/severe 1	
16. Unresolved issues	● late outcome after the Ross operation	

#### Postoperative valvar aortic stenosis

Criteria	Comments
1. Introduction and background	● common lesion ● most interventions in children are balloon dilation or open aortic valvotomy, aortic valve replacement, mechanical or biological prostheses or Ross procedure may have been performed
2. Survival – adult life	● excellent
3. Hemodynamic issues	● obstruction ● regurgitation ● LV function ● pulmonary homograft (Ross)
4. Arrhythmia/pacing	● arrhythmia rare ● more common in LV hypertrophy ● may cause sudden death

5. Investigations	ECG	<ul style="list-style-type: none"> <li>● routine LVH</li> <li>● conduction disturbances</li> <li>● repolarization changes</li> </ul>
	Chest X-ray	<ul style="list-style-type: none"> <li>● cardiomegaly</li> </ul>
	ECHO	<ul style="list-style-type: none"> <li>● see valvar aortic stenosis unoperated</li> <li>● prosthesis function and paravalvular leak</li> </ul>
	TOE	<ul style="list-style-type: none"> <li>● useful in assessment of paravalvular leaks and suspected endocarditis</li> </ul>
	MRI	<ul style="list-style-type: none"> <li>● rarely indicated</li> </ul>
	Cath	<ul style="list-style-type: none"> <li>● rarely indicated (see valvar aortic stenosis unoperated)</li> </ul>
	Exercise testing	<ul style="list-style-type: none"> <li>● surgical decision making for timing of reintervention</li> </ul>
6. Indications for reintervention	<ul style="list-style-type: none"> <li>● recurrent obstruction (native valve or prosthesis)</li> <li>● regurgitation</li> <li>● occasionally haemolysis</li> </ul>	
7. Interventional options	<ul style="list-style-type: none"> <li>● mechanical valve, homograft or Ross operation</li> <li>● prosthesis may be preferred by elderly</li> <li>● homograft may be preferred in endocarditis</li> </ul>	
8. Outcome	<ul style="list-style-type: none"> <li>● very good but anticagulant problems with mechanical valve and late failure</li> </ul>	
9. Endocarditis	<ul style="list-style-type: none"> <li>● prophylaxis in all cases</li> </ul>	
10. Pregnancy/contraception	<ul style="list-style-type: none"> <li>● anticoagulants may cause embryopathy</li> </ul>	
11. Recurrence/genetics/syndromes	<ul style="list-style-type: none"> <li>● see aortic valve stenosis unoperated</li> </ul>	
12. Physical activity/sports	<ul style="list-style-type: none"> <li>● high level activity possible in uncomplicated cases with good LV function</li> <li>● contact contra-indicated in patients on anticoagulants</li> </ul>	
13. Insurance	<ul style="list-style-type: none"> <li>● category 2</li> </ul>	
14. Follow-up interval	<ul style="list-style-type: none"> <li>● yearly</li> </ul>	
15. Follow-up care	<ul style="list-style-type: none"> <li>● Ross 1 otherwise 2</li> </ul>	
16. Unresolved issues	<ul style="list-style-type: none"> <li>● long-term outcome of Ross procedure</li> <li>● best anticoagulation protocol in pregnancy</li> </ul>	

## Subaortic stenosis unoperated

Criteria	Comments
1. Introduction and background	<ul style="list-style-type: none"> <li>● uncommon form of obstruction</li> <li>● may be discrete or extend to adjacent structures</li> <li>● often progressive.</li> </ul>
2. Survival → adult life	<ul style="list-style-type: none"> <li>● normal if obstruction not severe</li> </ul>
3. Hemodynamic issues	<ul style="list-style-type: none"> <li>● progression very common</li> <li>● may cause aortic regurgitation</li> <li>● associated lesions common (e.g. VSD)</li> </ul>
4. Arrhythmia/pacing	<ul style="list-style-type: none"> <li>● See 'aortic valve stenosis'</li> </ul>

5. Investigations	Chest X-ray	● baseline
	ECG	● routine LVH and repolarization changes
	ECHO	● investigation of choice ● visualise obstruction ● gradient across LV outflow tract ● LV mass/function ● aortic regurgitation
	TOE	● may be useful to define anatomy
	MRI	● rarely indicated
	Cath	● rarely indicated (see 'aortic valve stenosis')
	Exercise test	● for repolarization changes and symptoms
6. Indications for intervention	● progressive obstruction ● lower threshold and aortic valve stenosis ● aortic regurgitation	
7. Interventional options	● surgical resection	
8. Outcome	● recurrence possible	
9. Endocarditis	● prophylaxis in all	
10. Pregnancy/contraception	● low risk if no severe obstruction	
11. Recurrence/genetics/syndromes	● may occur left heart abnormalities e.g. coarctation, Shone's syndrome ● familial cases described	
12. Physical activity/sports	● no restriction if mild obstruction or after resection	
13. Insurance	● category 2	
14. Follow-up interval	● depends on severity and progression rate usually 1–2 yearly	
15. Follow-up care	● level 1	
16. Unresolved issues	● recurrence rate after resection ● optimal timing of surgery	

#### Unoperated coarctation

Criteria	Comments	
1. Introduction and background	● may present in infancy or later in adolescence	
2. Survival → adult life	● rarely undiagnosed in childhood but long term survival is possible	
3. Haemodynamic issues	● hypertension ● premature atherosclerosis ● LV hypertrophy/failure ● aortic dissection ● associated aortic/MV lesions	
4. Arrhythmia/pacing	● rare problems	
5. Investigations	ECG	● LVH repolarization changes
	Chest X-ray	● cardiomegaly ● ascending aorta dilation ● rib notching
	ECHO	● assessment of arch anatomy/gradient ● associated lesions LVH and function
	TOE	● rarely provides additional information
	MRI	● investigation of choice

	Holter	● not indicated unless for ambulatory blood pressure
	Exercise test	● hypertension on exercise ● arm/leg gradient ● inducible repolarization abnormalities
	Catheterization	● if MRI unavailable for arch anatomy ● for coronary angiography when indicated for intervention
	Additional	● screen for intracerebral vascular anomalies
6. Indications for intervention	● resting or exercise induced hypertension ● resting gradient >30 mmHg	
7. Interventional options	● balloon/stenting ● surgical repair	
8. Post treatment outcome	● residual hypertension common despite adequate relief of obstruction ● accelerated atherosclerosis ● reduced life expectancy	
9. Endocarditis	● prophylaxis in all cases	
10. Pregnancy/contraception/recurrence/fetal	● repair prior to pregnancy if possible ● transcatheter intervention may be indicated in unplanned pregnancy (worsening BP, LV failure) ● avoid oestrogen containing pill ● growth retardation common ● spontaneous fetal loss increased	
11. Recurrence/genetics	● recurrence may be familial ● 22q11 deletion in complex forms	
12. Syndromes	● Turners (present in approx 30%) ● Williams (present in approx 10%) ● Shones (associated LV inflow/outflow abnormalities)	
13. Sport/physical activity	● should be restricted prior to repair	
14. Insurance	● category 3 for significant unoperated coarctation	
15. Follow-up interval	● most patients referred for intervention on diagnosis ● 1 yearly of mild cases with BP at rest and exercise/ECHO/Doppler/MRI	
16. Follow-up care	● level 1	
17. Unresolved issues	● influence of age at operation on long-term outcome ● influence of drugs on vascular phenotype in successful cases ● role of intervention for mild gradients ● role of stenting as adjunct to balloon	

## Operated coarctation

Criteria	Comments	
1. Introduction and background		
2. Survival → adult life	● long-term survival still reduced despite adequate early repair	
3. Haemodynamic issues	● persistent and late developing hypertension at rest and exercise ● aortic valve dysfunction ● rare dissection	
4. Arrhythmia/pacing	● not an issue	
5. Investigations	EKG	● LVH±repolarization changes
	Chest X-ray	● cardiomegaly ● ascending aorta dilation ● rib notching
	ECHO	● assessment of arch anatomy/gradient ● associated lesions LVH and function
	TOE	● rarely provides additional information

	MRI	● investigation of choice
	Holter	● not indicated unless for ambulatory blood pressure
	Exercise test	● hypertension on exercise ● arm/leg gradient ● inducible repolarization abnormalities
	Catheterization	● if MRI unavailable for arch anatomy ● for coronary angiography when indicated for intervention
	Additional	● screen for intracerebral vascular anomalies advocated by some
6. Indications for intervention	● significant recoarctation (gradient >30 mmHg at rest) ● aortic aneurysm	
7. Interventional options	● balloon/stenting for anatomically suitable recoarctation ● surgery for complex situations±aneurysms	
8. Post treatment outcome	● excellent but late hypertension and premature atherosclerosis/CVA/MI/heart failure	
9. Endocarditis	● prophylaxis in all cases	
10. Pregnancy/contraception/recurrence/fetal	● relieve residual coarctation prior to pregnancy or during unplanned pregnancy ● monitor closely for hypertension ● avoid oestrogen containing pill if rest or exercise hypertension	
11. Recurrence/genetics	—	
12. Syndromes	—	
13. Sport/physical activity	● no restrictions if adequate relief of obstruction/no residual hypertension	
14. Insurance	● category 2	
15. Follow-up interval	● yearly with same investigations as for unoperated coarctation	
16. Follow-up care	● level 2	
17. Unresolved issues	● influence of age at repair, type of repair of intervention on late hypertension ● late outcome of balloon/stenting ● pathophysiology of late hypertension	

#### Patent arterial duct

Criteria	Comments	
1. Introduction and background		
2. Survival→adult life	● normal life expectancy in closed PDA ● rare PVD for large PDA	
3. Haemodynamic issues	● usually none – LV dilatation/pulmonary hypertension in significant PDA	
4. Arrhythmia/pacing	● none	
5. Investigations	Chest X-ray	● baseline ● cardiomegaly ● ductal calcification
	ECG	● usually normal ● LVH with large PDA
	ECHO/TOE	● usually diagnostic ● TOE rarely indicated

	Catheter	● for closure coronary angiography in older patients
	MRI	● not indicated
	Holter	● not indicated
	Exercise	● not indicated
	Additional investigations	● none
6. Indications for intervention	● controversial for silent or very small PDA ● continuous murmur ● LV dilatation	
7. Interventional options	● catheter closure Intervention of choice ● several device options ● surgery for rare cases	
8. Post treatment outcome	● excellent ● residual shunt in up to 10%	
9. Endocarditis	● not required after complete closure ● prophylaxis indicated otherwise	
10. Pregnancy/contraception/fetal	● no problems unless pulmonary vascular disease	
11. Recurrence/genetics	● none	
12. Syndromes	● congenital rubella	
13. Sport/physical activity	● no restrictions unless PVD	
14. Insurance	● category 1 for small PDA or after closure	
15. Follow-up interval	● discharge 1 year after closure	
16. Follow-up care	● level 3 unless PVD (1)	
17. Unresolved issues	● indication of closure for small PDA	

## Ebstein's anomaly

Criteria	Comments	
1. Introduction and background	● wide spectrum of pathologic anatomy which determines onset of severity of symptoms	
2. Survival → adult life	● extremely variable natural history ● infant survivors usually reach	
3. Haemodynamic issues	● cyanosis at rest and/or exercise (right – left shunt at atrial level) reduced exercise capacity ● congestive heart failure (tricuspid stenosis/ regurgitation/small RV) ● associated lesions ● LV abnormalities	
4. Arrhythmia/pacing	● atrial arrhythmias are common ● increase with age ● related to pre exultation and atrial dilatation ● risk of sudden death	
5. Investigations	Chest X-ray	● marked cardiomegaly ● right atrial enlargement
	EKG	● baseline (characteristic pattern) ● follow-up for rhythm
	ECHO/TOE	● severity of tricuspid valve displacement dysplasia and regurgitation ● RV size ● associated lesions ● LV function

	Catheter	● rarely required unless for coronary angiography in older patients or at EPS
	MRI	● rarely required
	Holter	● useful for arrhythmia monitoring
	Exercise	● baseline and follow-up ● cyanosis ● exercise tolerance ● arrhythmia
	Additional investigations	● EPS for arrhythmia diagnosis and RFA
6. Indications for intervention	● decrease in exercise tolerance ● heart failure ● increase in cyanosis ● arrhythmia	
7. Interventional options	● surgery for tricuspid valve repair or replacement ● RFA for arrhythmias/pre exultation	
8. Post treatment outcome	● symptomatic improvement usual ● tricuspid valve replacement – reoperation, thrombotic complications ● ongoing arrhythmia problems frequent ● risk of sudden death remains ● anticoagulants for atrial arrhythmia and prosthetic tricuspid valve	
9. Endocarditis	● prophylaxis in all cases	
10. Pregnancy/contraception/fetal	● well tolerated unless cyanosis or heart failure ● fetus at risk in cyanosed mother	
11. Recurrence/genetics	● 6% in affected mother. 1% in affected father. Familiar occurrence documented.	
12. Syndromes	● rare	
13. Sport/physical activity	● recreational sport in asymptomatic patient	
14. Insurance	● unoperated asymptomatic or well post operative category 2	
15. Follow-up interval	● depends on clinical status ● annual follow-up with ECHO/Holter/exercise test	
16. Follow-up care	● level 1 (operated and unoperated)	
17. Unresolved issues	● recurrence of arrhythmias ● long-term fate of repairs	

## Fontran

Criteria	Comments
1. Introduction and background	● palliative procedure for single ventricle physiology in which all systemic venous return directed to the lungs – multiple modifications
2. Survival → adult life	● improved survival with strict selection criteria ● late failure even in best cases
3. Haemodynamic issues	● function of systemic ventricle (preload deprived) ● pulmonary vascular resistance ● obstruction in Fontan connection ● atrial enlargement ● pulmonary venous obstruction ● AV valve regurgitation ● chronic venous hypertension ● desaturation/paradoxical embolus in fenestrated Fontan ● pulmonary arterio venous malformations in some

4. Arrhythmia/pacing	● atrial arrhythmias common ● increase with follow-up ● sinus node dysfunction ● pacing – ventricular pacing requires epicardial system	
5. Investigations	Chest X-ray:	● baseline and follow-up ● cardiomegaly ● pulmonary vascular markings
	ECG:	● rhythm
	ECHO/TOE:	● most useful investigation for ● ventricular function ● AV valve regurgitation ● residual shunts ● obstruction of Fontan connections ● thrombus in atrium ● routine TOE (2 yearly may be indicated or of arrhythmia present)
	Catheter:	● for haemodynamic assessment and angiography in clinical deterioration
	MRI:	● obstruction of Fontan connection ● occasionally useful for RA size and anastamieos
	Holter:	● routine and for symptomatic arrhythmia
	Exercise testing	● reaction activities only
	Additional investigations:	● blood/stool for PLE
6. Indications for intervention	● cyanosis ● obstruction to Fontan connection ● systemic AV valve regurgitation ● ventricular failure ● arrhythmia ● pulmonary venous obstruction	
7. Interventional options	● consider conversion to TCPC or transplant in failing Fontan ● closure of fenestration ● AV malformations ● RFA ● Supraventricular arrhythmia ● AV sequential pacing	
8. Post treatment outcome	● variable success with catheter ablation of atrial arrhythmias ● PLE has <50% 5 year survival ● Fontan conversion results unclearatrial arrhythmias common.	
9. Endocarditis	● prophylaxis in all	
10. Pregnancy/contraception/fetal	● pregnancy possible with perfectly selected patients and proper care ● high maternal risk in 'failing Fontan' ● higher miscarriage rate ● fetal risk of CHD may be higher ● avoid oestrogen pill if ejection fraction <40%, residual shunt, or spontaneous contrast in RA ● ACE inhibitors should be withdrawn if on anticoagulants – need meticulous management	
11. Recurrence/genetics	● non	
12. Syndromes	● none	
13. Sport/physical activity	● recreational sports only	
14. Insurance	● category 3	
15. Follow-up interval	● at least yearly review with ECHO, ECG Holter, exercise testing, blood testing	
16. Follow-up care	● level 1	
17. Unresolved issues	● indications for and results of Fontan conversion ● outcome of TCPC in modern era ● role of anticoagulation ● medical therapy for failing systemic ventricle ● role of ACE inhibitors	

## Marfan's syndrome

Criteria	Comments	
1. Introduction and background	<ul style="list-style-type: none"> <li>● abnormal fibrillin gene on chromosome 15q</li> <li>● autosomal dominant inheritance</li> <li>● cardiac defect largely determined outcome</li> </ul>	
2. Survival → adult life	<ul style="list-style-type: none"> <li>● death from cardiac problems</li> <li>● life expectancy reduced but improved by good cardiac follow-up and surgery</li> </ul>	
3. Haemodynamic issues	<ul style="list-style-type: none"> <li>● acute aortic dissection – risk higher if the aortic sinuses &gt;55mm</li> <li>● aortic regurgitation</li> <li>● mitral valve prolapse/regurgitation</li> </ul>	
4. Arrhythmia/pacing	<ul style="list-style-type: none"> <li>● atrial and ventricular arrhythmia in mitral valve prolapse/regurgitation</li> </ul>	
5. Investigations	Chest X-ray	<ul style="list-style-type: none"> <li>● not helpful for follow-up of aorta</li> </ul>
	ECG	<ul style="list-style-type: none"> <li>● rarely useful</li> </ul>
	ECHO/TOE	<ul style="list-style-type: none"> <li>● most valuable investigation for serial follow-up of aortic root dimensions, valve function (aortic and mitral)</li> </ul>
	Catheter	<ul style="list-style-type: none"> <li>● rarely indicated</li> </ul>
	MRI	<ul style="list-style-type: none"> <li>● excellent investigation for aortic arch and descending aorta</li> <li>● compliments echocardiography</li> </ul>
	Holter	<ul style="list-style-type: none"> <li>● not routine</li> </ul>
	Exercise testing	<ul style="list-style-type: none"> <li>● not routine</li> </ul>
	Additional investigations	<ul style="list-style-type: none"> <li>● non-cardiac assessment (ophthalmic, orthopaedic etc.)</li> </ul>
6. Indications for intervention	<ul style="list-style-type: none"> <li>● beta blockers for aortic dilatation</li> <li>● surgery if aortic diameter &gt;55 mm or rapid increase</li> <li>● significant aortic regurgitation</li> <li>● significant mitral regurgitation</li> </ul>	
7. Interventional options	<ul style="list-style-type: none"> <li>● urgent surgery for dissection</li> <li>● aortic root and valve replacement</li> <li>● valve sparing operation may be indicated</li> </ul>	
8. Post treatment outcome	<ul style="list-style-type: none"> <li>● Surgery improves life expectancy but other dissections still possible</li> <li>● beta blockers delay/prevent progression</li> </ul>	
9. Endocarditis	<ul style="list-style-type: none"> <li>● prophylaxis in valve regurgitation and after aortic surgery</li> </ul>	
10. Pregnancy/contraception/fetal	<ul style="list-style-type: none"> <li>● pregnancy contraindicated if aorta is &gt;45 mm</li> <li>● pregnant women should be on beta blockers</li> <li>● caesarean section to be discussed if aorta is dilated</li> </ul>	
11. Recurrence/genetics/syndrome	<ul style="list-style-type: none"> <li>● approximately 50% (autosomal dominant)</li> </ul>	
12. Sport/physical activity	<ul style="list-style-type: none"> <li>● strenuous exercise contra-indicated</li> <li>● high altitude and diving contra-indicated (spontaneous pneumothorax)</li> </ul>	
13. Insurance	<ul style="list-style-type: none"> <li>● category 3</li> </ul>	
14. Follow-up interval	<ul style="list-style-type: none"> <li>● annual follow up for aortic dilatation</li> <li>● more frequent evaluation if aortic diameter increasing</li> </ul>	
15. Follow-up care	<ul style="list-style-type: none"> <li>● level 1</li> </ul>	

16. Unresolved issues	<ul style="list-style-type: none"> <li>● role of early beta blockade</li> <li>● long term results of surgery including valve sparing</li> </ul>
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## Postoperative transposition (Mustard/Senning)

Criteria	Comments	
1. Introduction and background	<ul style="list-style-type: none"> <li>● common lesion – most Mustard/Sennings patients now adults – operation replaced by arterial switch mid 1980's</li> </ul>	
2. Survival → adult life	<ul style="list-style-type: none"> <li>● low early mortality</li> <li>● significant late morbidity/mortality from arrhythmia/baffle obstruction/RV failure with risk of sudden death</li> </ul>	
3. Haemodynamic issues	<ul style="list-style-type: none"> <li>● intra-atrial baffle obstruction (systemic and pulmonary venous) more common in Mustard than Senning</li> <li>● tricuspid regurgitation/RV failure relatively rare but important to detect early</li> </ul>	
4. Arrhythmia/pacing	<ul style="list-style-type: none"> <li>● progressive loss of sinus rhythm on Holter with follow-up</li> <li>● slow junctional rhythm may rarely require pacing</li> <li>● tachyarrhythmias (predominantly atrial flutter) may be related to high incidence of late sudden death</li> <li>● pacing may be required if antiarrhythmic drugs</li> </ul>	
5. Investigations	ECG	<ul style="list-style-type: none"> <li>● RVH with basic rhythm (often junctional)</li> </ul>
	Chest X-ray	<ul style="list-style-type: none"> <li>● useful for cardiomegaly</li> <li>● pulmonary venous obstruction</li> </ul>
	ECHO/TOE	<ul style="list-style-type: none"> <li>● TTE for ventricular function/tricuspid regurgitation</li> <li>● TOE essential if questions remain regarding baffle function</li> </ul>
	MRI	<ul style="list-style-type: none"> <li>● rarely required if TOE available</li> </ul>
	Holter	<ul style="list-style-type: none"> <li>● occult arrhythmia</li> <li>● not predictive of SD</li> </ul>
	Exercise test	<ul style="list-style-type: none"> <li>● exercise tolerance</li> <li>● evaluation of arrhythmia</li> </ul>
	Catheterization	<ul style="list-style-type: none"> <li>● for intervention and assessment of new onset symptoms</li> </ul>
	Additional	<ul style="list-style-type: none"> <li>● EP study/RFA for refractory atrial arrhythmias</li> </ul>
6. Indications for intervention	<ul style="list-style-type: none"> <li>● baffle obstruction</li> <li>● baffle leaks</li> <li>● tricuspid valve dysfunction</li> <li>● RV failure</li> </ul>	
7. Interventional options	<ul style="list-style-type: none"> <li>● balloon/stenting for pathway obstruction</li> <li>● transcatheter closure for baffle leaks</li> <li>● tricuspid valve/replacement</li> <li>● conversion to arterial switch (pulmonary artery banding)</li> <li>● transplantation</li> </ul>	
8. Post treatment outcome	<ul style="list-style-type: none"> <li>● risk of sudden death despite lack of symptoms or overt haemodynamic disturbance</li> </ul>	
9. Endocarditis	<ul style="list-style-type: none"> <li>● prophylaxis in all cases</li> </ul>	
10. Pregnancy/contraception/recurrence/fetal	<ul style="list-style-type: none"> <li>● pregnancy not contra-indicated in most cases</li> <li>● monitor RV function throughout</li> <li>● no contraceptive issues</li> <li>● long-term consequences on RV function not known</li> </ul>	
11. Recurrence/genetics	<ul style="list-style-type: none"> <li>● familial recurrence of TGA rare</li> </ul>	
12. Syndromes	<ul style="list-style-type: none"> <li>● none</li> </ul>	

13. Sport/physical activity	● generally normal activities ● maximal exercise tolerance likely to be diminished
14. Insurance	● category 3
15. Follow-up interval	● yearly
16. Follow-up care	● level 1
17. Unresolved issues	● risk stratification for sudden death ● fate of systemic RV/tricuspid valve ● indication/conversion/transplant strategies

### Congenitally corrected transposition

Criteria	Comments	
1. Introduction and background	● rare lesion ● usually associated with other abnormalities ● may occur with dextrocardia	
2. Survival → adult life	● common to survive to adult life ● associated lesions common (VSD, PS, left AV valve regurgitation) determine outcome	
3. Haemodynamic issues	● cyanosis with VSD and PS ● PVD if VSD and no PS ● systemic ventricular failure with systemic A-V valve regurgitation ● referral before systemic ventricular dysfunction	
4. Arrhythmia/pacing	● spontaneous CHB (2% per year) and post surgical heart block ● endocardial pacing in the morphologic LV ● atrial arrhythmias common ● ventricular arrhythmias with systemic ventricular dysfunction ● epicardial pacing if potential for paradoxical embolus	
5. Investigations	Chest X-ray:	● baseline ● follow-up for associated lesions ● cardiomegaly
	ECG:	● rhythm
	ECHO/TOE:	● size and function of systemic ventricle ● morphology of left A-V valve ● associated lesions
	Catheter:	● for pulmonary haemodynamics and anatomy of associated lesions
	MRI:	● rarely required
	Holter:	● for occult arrhythmia detection
	Exercise function:	● helpful for timing of surgery ● oximetry ● exercise tolerance
	Additional investigations:	● occasionally MUGA for ventricular function
6. Indications for Intervention	● +>moderate systemic AV valve regurgitation ● significant associated lesions ● pacemaker for complete AV block with symptoms, profound bradycardia or chronotropic incompetence	
7. Interventional options	● valve replacement ● pulmonary artery banding ● 'double switch' (controversial in adults)	
8. Post treatment outcome	● good if left A-V valve replacement before systemic ventricular function deteriorates ● atrial arrhythmias common	
9. Endocarditis	● prophylaxis in all cases	

10. Pregnancy/contraception/fetal	<ul style="list-style-type: none"> <li>● pregnancy not contra-indicated if asymptomatic</li> <li>● monitor ventricular function and rhythm</li> <li>● long-term consequences on systemic ventricular function unknown</li> <li>● avoid oestrogen containing contraceptive pill if cyanosed/pulmonary hypertension</li> </ul>
11. Recurrence/genetics	<ul style="list-style-type: none"> <li>● 4%</li> </ul>
12. Syndromes	<ul style="list-style-type: none"> <li>● none</li> </ul>
13. Sport/physical activity	<ul style="list-style-type: none"> <li>● no restriction on recreational activities</li> </ul>
14. Insurance	<ul style="list-style-type: none"> <li>● Category 3 in most cases</li> </ul>
15. Follow-up interval	<ul style="list-style-type: none"> <li>● yearly, with ECHO, exercise test±Holter</li> </ul>
16. Follow-up care	<ul style="list-style-type: none"> <li>● level 1 (pre and postoperative)</li> </ul>
17. Unresolved issues	<ul style="list-style-type: none"> <li>● classical repair of VSD and PS versus 'double switch.'</li> </ul>

## 8 Recommendations for Future Developments in Europe

### 8.1 Summary

This task force seeks to describe the increasing population of grown-ups with congenital heart disease, promote a review of their needs and make proposals for service delivery. The representatives were chosen from Europe, Canada and United States because they had pioneered the subject in their countries, as well as contributed to previous guideline documents. It is recognized that this is a 'speciality in evolution' and that new information is likely to be acquired rapidly. There is an urgent need to establish a comprehensive hierarchical service with dedicated specialist units and defined referral links. This report proposes a framework, which should be adapted by health care systems in European countries and developed further. Cases of congenital heart disease in adults will strongly outnumber those in children and the number will continue to grow. We hope the contents of this report will be a stimulus to investment to enable patients to be managed in adequately staffed and funded expert units and to continue to receive the excellent level of health care provided to them during childhood.

### 8.2 Specific recommendations

- 1 Patients should be transferred to appropriate adult care at a flexible age of 16–18 years.
- 2 There should be a transitional service for 12–16 year olds to facilitate this process.
- 3 Each paediatric cardiology and adult cardiology unit should have a defined referral path into the grown-up congenital heart disease service.
- 4 A hierarchical system for care delivery should be co-ordinated by specialist regional units for grown-ups with congenital heart disease.
- 5 These regional units should be sited within adult cardiology programmes in multidisciplinary teaching environments.
- 6 The specialist units should be fully staffed and resourced, to provide optimal investigation and treatment for patients with complex congenital heart disease.
- 7 After definition of level of care required at the specialist unit, patients should be looked after, either exclusively in the specialist unit (level 1), have shared care between the specialist unit and trained local cardiology units (level 2) or be followed in their local unit (level 3).
- 8 Follow up plans should be clearly communicated to the patient, their primary care physician and local cardiology units. Patients should be provided with a 'health passport' containing key information on their condition, treatment, and outcome issues and follow-up plan.
- 9 Training programmes for specialist staff and cardiologists with an 'interest' should be defined and implemented.
- 10 Most specialist examinations, interventional catheterizations and surgical procedures should be performed at the specialist centres.
- 11 The specialist centres should provide the framework for training and the establishment of a comprehensive national database for research.
- 12 The specialist centres' staff should have expertise in medical and psychosocial management.
- 13 As far, as possible, the provision of health care delivery for grown-ups with congenital heart disease and training of staff should be standardized and co-ordinated through the European Society of Cardiology.

## 9 European Society of Cardiology Staff

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### Appendix A

#### Adult congenital heart disease survival simulator

Thank you for downloading this simple simulation program. It is intended to model the rate of attrition of a population, such as a particular cardiac condition for which cardiac surgery has been performed.

Because surgery has improved considerably two phases for which different mortality rates can be selected have been incorporated. You need to select a death rate per year (percent) and the length of the first phase, and then the death rate for the second phase, and the total length of follow up for both phases.

This will run in Microsoft Excel. Because there are modules of automated code, when loading the file, Excel will alert the user that the file contains Macros. This should not concern the user, but it does not have to be said that every computer user must take responsibility for using an up to date virus checker. The programme was free of virus according to the most up to date McAfee Virus Scan, but no responsibility can be taken during further distribution.

This is a non-commercial program and must not be redistributed for profit. Full copyright remains with Dr Graham Derrick (grahamderrick@doctors.org.uk). The source code can be made available to those who request it, with the request that any modifications are returned to Dr Derrick.

This simulation program can be consulted and downloaded from the ESC web site at the following address: [www.escardio.org](http://www.escardio.org), in the Scientific & Clinical Information/Guidelines & Scientific Statements/Task Force Guidelines' section.

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