CONGENITAL HEART DISEASE IN ADULTS:
NEW PROBLEMS FOR THE CARDIOLOGIST

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THE SUBJECT OF GROWN-UP congenital heart disease (CHD) is currently and correctly attracting attention in the world.

Surgical treatment for congenital heart disease (CHD) has evolved through the last 40 years starting with the attempts by Blalock and Brock to palliate, to the current refined "technical virtuoso" repairs of the most complex problems during infancy. The explosive development of pediatric cardiology as a speciality has been inevitable, reaching maturity in 1980 with its own World Congress. In 1976 a unit for adolescent cardiac medical and surgical cases was opened in the National Heart Hospital (NHH), London, when the increasing and continuing demands of the young cardiac survivors with congenital malformations who had been treated in infancy and childhood were recognized. The provision of vital services for the transition between pediatric and adult cardiac medicine and surgery has become an integral part of the GUCH (grown-up congenital heart) services to adolescents and adults with congenital heart problems.

Although the British Heart Foundation suggests in its 1989 advertisement that 90% of patients born with congenital heart disease survive to adulthood, calculations based on many assumptions suggest that the correct figure is closer to 70%.

There has been a steady increase in adolescent cardiac admissions (aged 12 to 19 years), particularly since union in 1989/90 with the Brompton, with its active pediatric cardiology department. A total of 95% of referred young patients have CHD, 75% of admissions having had earlier palliative and/or reparative surgery. Predictably, in the last seven years, GUCH admissions over 20 years and 30 years exceed the adolescents as the population increases and ages. Thus, the problem is not in provision for adolescents during the transition period alone, it is for a larger, more difficult group of adults. Even though many pediatric cardiologists consider it is their right to continue the care of GUCH, it is not a pediatric problem and most adolescents resent or refuse treatment within the settings for children or to be treated as children. It does not matter whether pediatric or adult cardiologists are responsible for GUCH. What does matter is that they are educated and acquire the necessary special skills and knowledge, i.e., train as in any other speciality of cardiology such as intervention, electrophysiology, etc. The medical demands from adolescent cardiac patients in the 1970s and 80s are the same but differ in incidence, reflecting the changes in the 1970s and advances in technology and therapy (Figure 1). Better imaging by echocardiography, including transesophageal, and magnetic resonance imaging (MRI), has diminished the need for invasive investigation but an equivalent increase in laboratory time and skill is needed for therapeutic interventions. Provision of day case facilities in 1984 diminished the admission requirements as patients come from long distances for such specialized services. For the GUCH unit to be complete, ideally it should be both geographically close to pediatric cardiology for easy referral and transitional care problems with adolescents and closely integrated with cardiac surgery — as in the Royal Brompton. The services needed for ideal GUCH care are summarized in Table 1.

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Figure I. The changing reasons for admission of adolescents' young adults with congenital heart disease in the 1970s and the 1980s.

Table 1. Skills and technology required for GUCH management.

- Adult cardiology - trained in congenital heart disease
- Pediatric cardiology (on site ideal)
- Cardiac surgery with expertise in GUCH
- Intensive care with expertise in GUCH
- Adolescent unit
- Day case facilities
- Magnetic resonance imaging
- Transesophageal echocardiography
- Cardiac catheter laboratory for congenital heart disease
- Biplane angiography
- Interventional procedures for GUCH
- Complex arrhythmia investigation and pacemaking
- Transplant = heart and lung
- Special "at-risk" pregnancy clinic

Medical (Cardiology) Demand for Inpatient Services

Arrhythmias

This is the commonest reason for admission in all decades, becoming more frequent with increasing age. Arrhythmias occur as part of the natural history of many cardiac anomalies. Cardiac surgery may delay onset or induce the arrhythmia in the natural history or add new ones prematurely. Usually, a symptomatic deterioration occurs from paroxysmal and/or established rhythm disorders, thus requiring identification and suppression and correct management of the underlying cardiac problem, something the adult cardiologist often ignores to the detriment of the patient's well-being. The need to investigate and prevent sudden unexpected death is demanding on resources and still unsuccessful. Cardiologists, who rarely understand the underlying disorder, tend to ignore it and often use drugs which cause accelerated deterioration in the function of the already abnormal myocardium. Informed help tends to be sought only when disaster and irreversible problems have occurred. There is a temptation to control the rate of atrial flutter and fibrillation with drugs such as amiodarone and beta-blockers and ignore the pressing need to restore sinus rhythm. There is a disquieting singleminded obsession to investigate and master ventricular arrhythmias as a cause of sudden death and ignore the role of atrial flutter. Atrial flutter is the commonest arrhythmia post-Fontan, in single ventricle, tricuspid atresia, and post-Mustard. It occurs often in Fallot, associated with right ventricular dysfunction and worsening it, and often is related to malfunctioning conduits or chronic pulmonary regurgitation in native valves. Three aborted "sudden" deaths in this unit's series have been shown to be caused by atrial flutter, having been treated with the wrong drug on the erroneous assumption that ventricular tachycardia was the cause. Sophistication is needed to differentiate atrial flutter with a one-to-one response in a patient with right wide bundle branch block, so frequently present in post-surgical heart disease, from ventricular tachycardia. The adult cardiologist's knowledge and wisdom relating to management of arrhythmias must be united with the pediatric cardiologist's understanding of anatomy and postoperative complications. Too often the danger of the thromboembolic complication is ignored by pediatricians and thrombus often collects in the diseased fibrillating right atrium - a particular hazard in Fontan where the right atrium can become paralyzed and allow stagnant flow within. The importance of looking for obstruction in the Fontan circulation is constantly ignored by the adult cardiologist.

Serious problems are often encountered by patients needing pacemakers and recurrent changes, which should be performed by experts, not by junior staff in training. There is often a
shortage of venous access, abnormality of drainage, and atrial anatomy. Profound and permanent functional deterioration may occur in complex hearts after pacemaker failures. Sepsis in pacemakers has caused death in GUCH patients. Patients with heart block complicating their lesions have a worse long-term outcome than their counterparts with sinus rhythm, even without pacemaker failure. This highlights the special importance and risks of pacing in GUCH and confirms the need for the expert.

Endocarditis

Despite instructions to patients and doctors, endocarditis occurs in postoperative congenital heart disease (Table 2) as well as unoperated CHD and there is considerable delay in diagnosis. Some lesions are not affected, thus prophylaxis is unnecessary in these (Table 3) as is consideration of the diagnosis. This does not exclude secondary involvement of the cardiac lesion in a generalized septicemia from coxiella, staphylococcus aureus, brucella, etc. Doctor and patient are aware of the risks of endocarditis and may delay with prophylaxis but doctors fail to consider the possibility of the diagnosis, prescribe antibiotics, and fail to do appropriate blood tests or referral.

Table 2. Incidence of endocarditis in 47 patients aged 6-30 years with operated congenital heart disease.

<table>
<thead>
<tr>
<th>Left valves</th>
<th>65%</th>
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<tbody>
<tr>
<td>T.G.A. (Rastelli) V.S.D.</td>
<td>4</td>
</tr>
<tr>
<td>(Open)</td>
<td>6</td>
</tr>
<tr>
<td>Fallot</td>
<td>8</td>
</tr>
<tr>
<td>Complex repairs</td>
<td>9</td>
</tr>
<tr>
<td>Cont. Intra-Op. Infection</td>
<td>8</td>
</tr>
</tbody>
</table>

V.S.D. = ventricular septal defect; T.G.A. = transposition of great arteries.

Table 3. Postoperative congenital heart lesions that do not require protection against bacterial endocarditis.

Simple pulmonary valvotomy
Secundum atrial septal defect (without mitral valve lesion)
Ventricular septal defect (completely closed without left valve lesion)
Total anomalous pulmonary venous drainage
Duct with no residual valve lesion
Eisenmenger patients without valve abnormalities

before giving the treatment for an undiagnosed condition. The result is ODD (delay and destruction and disaster). The dangers of not making an early diagnosis and therefore suppressing it by inadvisable therapy cannot be over-emphasized. It should be regarded as negligence. The only hope is to educate the patients not to take random antibiotics before appropriate blood tests which must include blood cultures.

Terminal and Transplant Assessment

Myocardial failure may result from residual lesions, damage to the myocardium sustained during early long bypasses, or from congenital abnormality of the myocardium as part of diffuse congenital cardiovascular disease over and above the compensatory hypertrophy and dilatation and acquired myocardial dysfunction, cause unknown. Prior to 1975, myocardial protection during bypass was inadequate, damage occurred from potassium arrests, bad technique and long operations without adequate or, sometimes, any coronary perfusion. When patients survived, healing fibrosis and even excessive endocardial fibrosis looking like fibroelastosis can occur as late sequelae, probably the result of subendocardial ischemia during bypass, now uncommon. However, in a specialized unit young patients in terminal failure collect, waiting and hoping for heart or heart-lung transplant. Such patients use many resources and as a result of shortages in finance and personnel may prevent less hopeless routine problems being solved. Patients are accepted for transplant which they may never get owing to the shortage of donors; emotional and costly medical support must be provided by the unit. Probably 85% of early Mustards, many Fontan patients, most univentricular hearts, and many other malformations will need transplantation before age 30 years if survival is to be prolonged.

Surgical Demands

Cardiac surgery is needed in the GUCH community. The type of surgery has changed over the last decade with increasing needs for reoperation on the definitive repairs and radical palliations. With improved and increased direct
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repairs during infancy and childhood, there are fewer GUCH patients presenting for first operations. Reoperations are mainly for degeneration in valves and conduits; this involves most (?all) patients who have had "repair" using conduits and valves. Endocarditis and new lesions may require reoperation. The morbidity and mortality may be high in certain conditions (Table 4). Parents and patients should be warned of the need in the future and not be dispatched during childhood with the belief that they are "cured" for all time. Many problems occur at "re-opening", some catastrophic, hence the need for the experienced surgeon, not the enthusiastic junior. Cardiac surgical expertise in the older congenitals will be needed for the inevitable acquired coronary artery disease in the elderly GUCH patients, but is very rare in cyanotics' who have large coronary arteries.

A problem for GUCH cardiac surgeons is the delay by cardiologists in referring patients back for surgery before serious deterioration in myocardial function occurs - this is an important contributor to mortality. Right -sided obstruction does not usually produce symptoms until serious and sometimes irreversible failure occurs. Recognition of important obstruction is not easy in this setting and requires understanding of subtleties, like the management of arrhythmias and the need and interpretation of magnetic resonance imaging. Unfortunately, cardiologists often fail to seek expert help. They forget the importance of reading the original operation report and they do not know the natural history of the various valves or conduits. The mortality for "redo" Fontan in GUCH is currently very high.

Undoubtedly, this is contributed to by original errors of selection and technique, as well as late referral; some of the mortality can be avoided but this figure suggests that there is a problem which other units should recognize when they are undertaking both the first and subsequent operations. The urgent need for research into preventing or delaying calcification of biological valves cannot be over emphasized.

As techniques have improved, the residual and late problems have changed. Today's problems will not be tomorrow's, but cardiac surgical expertise is and will be needed for grown-up CHD. How, where, and by whom with what training requires to be addressed by the cardiac surgical hierarchy.

### Outpatient Support Services

There are increasing outpatient referrals and problems as each decade's survivors are added to the last. Besides the cardiac problems, patients, both well and ill, seek advice about normal life's activities and demands in relation to their heart condition (Table 5). It is important that such advice is given by the informed since poor management may contribute to loss of life or function. The general surgeon often fails to seek advice and the physician is called too late after unnecessary complications. The plastic surgeon may be needed to even the incised breast or reduce deforming scars. Health insurance for these patients is a problem but important with the decline of National Health Services. Life insurance companies in the United Kingdom appear disinterested unless the sums are large and

| Table 5. "Left" problems faced by GUCH patients in relation to their cardiac condition. |

| Insurance |
| Pregnancy |
| Risk of affected offspring |
| Driving licences (ordinary and vocational) |
| Employment |
| Housing priority |
| Social service support |
| Contraception |
| Prison management and sentences |
| Psychological problems in relation to their heart disease |

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Table 4. Reoperation age 12 years and over for congenital heart disease. First re-operation on definitive repair.

<table>
<thead>
<tr>
<th></th>
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<tbody>
<tr>
<td>Aortic valve stenosis</td>
<td>59</td>
<td>6(10%)</td>
</tr>
<tr>
<td>Fallot</td>
<td>26</td>
<td>3(12%)</td>
</tr>
<tr>
<td>Right ventricular outflow Tract reconstruction</td>
<td>26</td>
<td>2 (8%)</td>
</tr>
<tr>
<td>Fontan</td>
<td>18</td>
<td>8(44%)</td>
</tr>
<tr>
<td>Ventricular septal defect + Aortic regurgitation</td>
<td>17</td>
<td>4(24%)</td>
</tr>
<tr>
<td>Subaortic stenosis</td>
<td>13</td>
<td>2(15%)</td>
</tr>
<tr>
<td>Coarctation</td>
<td>13</td>
<td>0</td>
</tr>
</tbody>
</table>

then the patient is refused or has to pay excessively high rates. Support clinics for "at-risk" pregnancy, transesophageal echocardiography, fetal scanning, and pre/post transplant are needed once a service for grown-up CHD is established. Support for the bereaved GUCH family is needed from the GUCH cardiologist as families may return to the GUCH unit for many years after the death.

Who Knows? Who Cares?

The organization for long-term care of GUCH patients is required now in any country which has had effective pediatric cardiac surgery for infants and children over the last 20 years. "Who knows", like "who cares" is not agreed. Many think they know. Few recognize to know is necessary. Even fewer actually know. Many patients return to the surgeon whose name they remember, unlike that of the pediatric cardiologist! However, the surgeon's experience and attitude to late problems relate to mortality and reoperation with the frequent assumption that the rest are "well". It is not. The pediatric cardiologist is familiar with patients and the underlying anomaly but unused to the one-to-one relationship needed for adult patients, particularly the separation from mother so necessary in adolescents; the different cardiac problems encountered as an adult; the adult drug regimes, and the management of coronary heart disease and hypertension which will be required by some. An additional difficulty is that most pediatric cardiologists do not have access and/or control of adult inpatient beds, particularly when coming from the children's hospital. The local cardiologist, to whom these patients are usually referred by general practitioners and physicians, is often ignorant of the underlying condition and its complications and frightened of its nomenclature, see only the management of rhythm disorders and failure in routine cardiological terms, and tend to refer patients back for operations too late and with a reluctance (?arrogance) to seek help from the few informed sources. GUCH patients hopefully will spend most of their lives as adults and need adult-trained doctors. This is not to say pediatric cardiologists cannot do this, they can, but they must have adequate training. After all, the pediatricians in most countries, correctly, have invested much time and effort in preventing adult doctors looking after infants and children because of the lack of special skills and facilities. So, the converse is true. To improve the care of the GUCH, the subject should be one of the subspecialities of cardiology, like electrophysiology, intervention, or echocardiography. The methods and organization of GUCH care must be flexible because of the different medical, social, and welfare environment of the patients. Special education of a few cardiologists and enthusiasm are vital, as well as the recognition that in most countries these patients are getting inadequate care. Some pediatric cardiologists already have enough experience in adult cardiology in their training and practice (recommended in the United Kingdom as a minimum of six months for the training of a pediatric cardiologist): they must organize adequate inpatient facilities and integrate with the department of cardiology.

The management of GUCH needs a number of basic skills, including knowledge of prognosis and well-being of patients after the first decade of follow-up. For this, an Ability Index1 has been created (Figure 2). The index has taken into account the patient's perception of normality of

**ABILITY INDEX**

1. Normal life. Full time work or school. Manage pregnancy.
3. Unable to work. Limitation all activities. pregnancy risk.
5. Socio/Community Imposition because CARDIAC ANOMALY

Figure 2. Ability Index classification to define the capability of patients to lead a normal or restricted life, taking into account their adaptation to having or having had congenital heart disease.
life, as well as the doctor's! It is not based on exercise tolerance and thus has more relevance than the New York Heart Association classification. For ease of lecturing, it has been color coded by the readily remembered colors of the rainbow and is thus useful for quick comparison of results, saving verbose description.

Acquiring the skills needs training and fundamental knowledge of the incidence and the particular complications of each lesion and operation. This knowledge is needed for counselling, prevention of problems, and advice on way of life for the young. The incidence of late serious problems differs according to diseases and procedures (Figures 3A and 3B); the surgeon is a rarely considered factor, but is important. One unit's results are not the same as another's, nor are the late complications. With experience, one can often tell where a patient with Fallot's tetralogy has had repair by the residual lesions and late problems!

### Figures 3A & B: Incidence of important post-operative problems in various congenital heart lesions.

#### Organization

Integrated with the proper provision of medical expertise is the basic organization and support services for long-term care of the grown-up CHD patient. The ROCK concept has been designed for this: (1) Recognise the problem – pediatric cardiologist, parent and patient take responsibility; (2) Organize referral where there is or will be expertise. This must be flexible according to where the patient has come from, i.e., special cardiac center, children's hospital or university clinic. Pediatric cardiologists must be the major people responsible for this organization and referral. They may chose and/or train interested cardiological colleagues or do joint clinics. It must be decided and known where all original notes will be for the patient's life. The current policy of note destruction in many centers of the United Kingdom (including our own), particularly if patients are not seen for 10 years, is jeopardizing optimal care; (3) Centralise the specialist needs and expensive technology in a few centers, i.e., 4 to 5 for the United Kingdom. Facilities should not be diluted by every cardiac center wanting to "have a go". There are relatively few patients and expertise, particularly in the tough problems, will be hard to acquire if there is dissipation and small numbers; and (4) Knowledge and skills required - available and centralized (Table 1).

Currently in Europe, there are few countries organized correctly, but more centers are being established. There are few trained cardiologists/pediatric cardiologists with the appropriate knowledge. Some from each speciality take up GUCH care without "training" or exposure but out of need. A survey done through the members of the Association of European Pediatric Cardiologists confirms this state of chaos. However, judging from the plenary sessions and "state-of-the-art" lectures sought, European cardiology as well as Japan and the United States are gradually stirring and their awareness must be the start.

#### Conclusion

As can be seen there is much work to be done for the "grown-up" congenital heart patients. This
new medical community has evolved from the dramatic progress of medicine and surgery. The welfare and needs of the patient (not status and wishes of individuals, physicians or organizations) must be the first consideration. Medical society has already lavished expert care, technology, and finance to ensure their survival.

Cardiologists must accept the new responsibility for the care of the GUCH, providing the correctly trained experts and education of the doctors and health care authorities about their specialist needs and costs. Medical care cannot be relinquished once the pretty blue baby becomes an unattractive adult. The GUCH deserve our professional best and long-term concern.