Adults with Congenital Heart Disease: Multiple Needs of a Fast Growing Cardiac Patient Group

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Congenital heart disease (CHD) is one of the most common inborn defects, occurring in approximately 0.8% of newborn infants. Adults with congenital heart disease are the beneficiaries of successful paediatric cardiac surgery and paediatric cardiology programmes across the developed world. Fifty per cent or more of them would have died before reaching adulthood had it not been for surgical intervention in infancy and childhood. This dramatic success story has created a large and growing population of young adults, who require life-long cardiac and non-cardiac specialised services[1].

It is now well appreciated that most patients with CHD who have had their lives transformed by surgical intervention(s) had reparative and not corrective surgery (Figure 1)[2]. Many of them face the prospect of further surgery, arrhythmia[3] and other complications[4] and, if managed inappropriately, are at increased risk of heart failure and premature death[5]. There are approximately 800.000 adults with CHD in the USA[6], 200.000 in the UK and 30.000 patients in Greece. These numbers will continue to grow as more and more children become adults. With current advances in cardiac surgery, peri-operative care and our better understanding of CHD, more than 85% of infants are expected to reach adulthood. A 400% increase in adult outpatient clinic workload was recently reported in Canada (Figure 2)[7]. In the UK, the need for follow-up of patients with moderate to severe complexity CHD over the age of 16 years has recently been estimated at 1600 new cases per year[8]. Furthermore, there are patients with structural and/or valvular congenital heart disease who present late during adulthood[9]. Most of these patients will both require and benefit from expert cardiology care.

In general, attendance at a regional adult care centre should be considered for:

a) the initial assessment of suspected or known CHD,

b) follow up and continuing care of patients with moderately severe and complex lesions,

c) further surgical and non-surgical intervention and,

d) risk assessment and support for non-cardiac surgery and pregnancy.

The majority of adults with congenital heart disease will still require local follow-up for geographic, social, and/or health economic reasons. Primary care physicians and general adult cardiologists must, therefore, have some understanding of the health needs and special issues in the general medical management of this relatively new adult patient population. Importantly, community and hospital physicians must recognise when to promptly refer these patients to an expert centre. Published management guidelines[8] may assist in this process[10].

A new set of recommendations have been made following the 2001 Bethesda

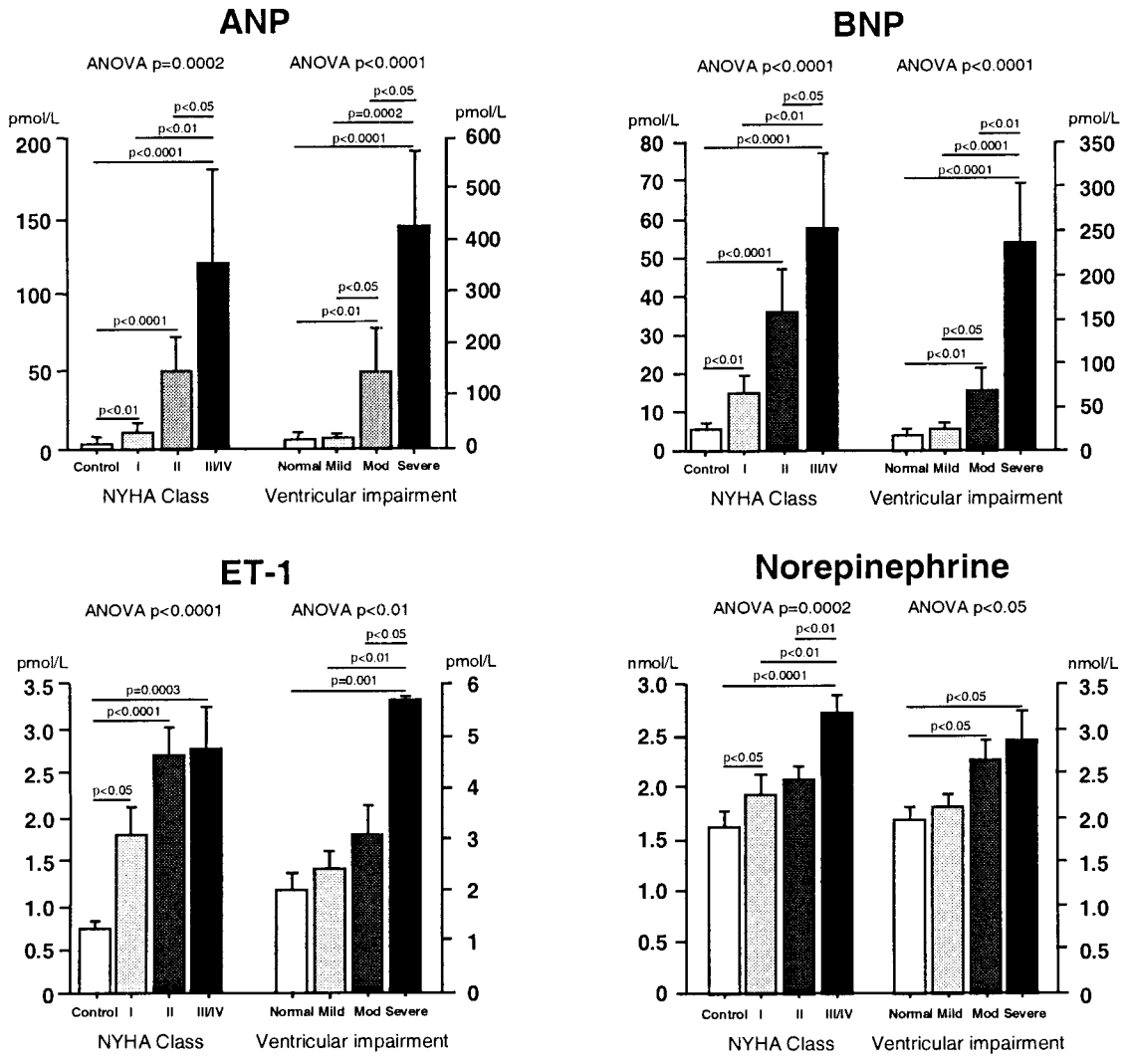

Figure 1. Neurohormonal activation and the chronic heart failure syndrome in adults with congenital heart disease. Neurohormone levels in 53 adult patients with congenital heart disease attending the Royal Brompton Outpatient Clinic; relations to New York Heart Association (NYHA) functional class and to systemic ventricular function (note differing y-scales for each measure). ANP: atrial natriuretic peptide, BNP: brain natriuretic peptide, ET-1: endothelin-1. *(With permission from Bolger et al, Circulation 2002; 106: 92-99).*

Conference[11] regarding care delivery systems, improved access to health care, manpower planning and training objectives.

Organization of care

Care of the adult with CHD should be co-ordinated by regional or national adult CHD centres, fulfilling the following purposes:

- To optimise care for all adult patients with CHD and to reduce errors in their care.
- To consolidate specialised resources required for the care of adult congenital heart patients.
- To provide sufficient patient clinical volumes to facilitate specialist training for medical and non-medical personnel, and to maintain staff and faculty competence and special skills in the treatment of adult patients with CHD[12].
- To facilitate research in this unique population, work towards the ideal of evidence-based care, and promote a more complete understanding of the late pathophysiology and determinants of late outcomes in these patients.
- To offer educational opportunities and continuous support to primary caregivers, cardiolo-

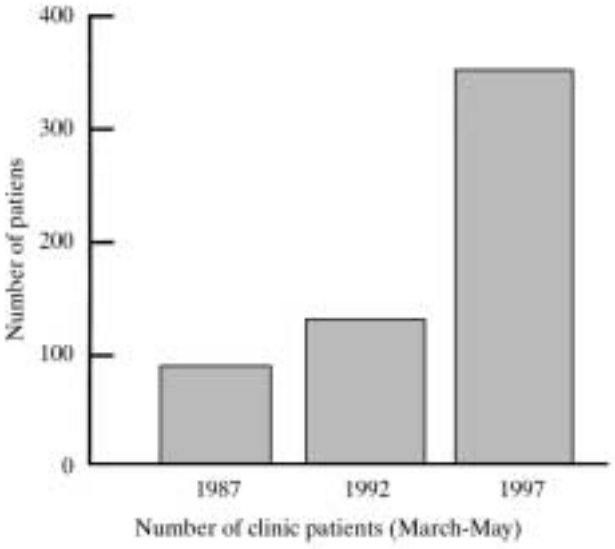

Figure 2. Increasing workload of outpatient clinics for adults with congenital heart disease.
Patients seen at the TCCCA clinic, Toronto, Ontario, Canada during the same three month period in 1987, 1992 and 1997. Notice 44% and 269% increases between 1987 and 1992 and 1992 and 1997, respectively.
(With permission from Gatzoulis et al, Heart 1999; 81: 57-61).

gists and surgeons so that they may contribute optimally to patient management.

- To provide a readily available source of information and expert opinion for patients and doctors.
- To help organise support groups for patients.
- To provide information for government and act as the representative of the speciality.

Approximately one regional or expert centre should be created to serve a population of 5-10 million people. Greece, therefore, with a population of approximately 10.000.000 people should ultimately support two such adult CHD centers.

- Adults with moderate and complex CHD (Table 1) will require periodic evaluation at a regional adult CHD centre. These patients, as indeed all patients with CHD, should also and will also benefit from maintaining regular contact with their primary care physician (in a joint care model).
- Existing paediatric cardiology programs should identify or help develop an adult CHD centre to which transfer of care should be made when patients reach adult age.
- Similarly, adult cardiology and cardiac surgical centres and community cardiologists should have a referral relationship with a regional adult CHD centre.
- All emergency care facilities should have an affiliation with a regional adult CHD centre.

Table 1. Types of patients who should be seen at regional adult CHD centres *(alphabetically)*.

Absent pulmonary valve syndrome
Aorto-pulmonary window
Atrioventricular septal defects
Cardiac tumours
Coarctation of the aorta
Common arterial trunk (or Truncus arteriosus)
Congenitally corrected transposition of the great arteries
Cor triatriatum
Coronary artery anomalies (except incidental findings)
Criss-cross heart
Cyanotic congenital heart patients (All)
Double outlet ventricle
Double inlet ventricle
Ebstein anomaly
Eisenmenger syndrome
Fontan procedure
Interrupted aortic arch
Isomerism (Heterotaxy syndromes)
Kawasaki's disease
Infundibular right ventricular outflow obstruction (moderate to severe)
Marfan syndrome (unless already established under expert leadership)
Mitral atresia
Single ventricle (also called double inlet, double outlet, common/primitive ventricle)
Partial anomalous pulmonary venous connection
Patent ductus arteriosus (not closed)
Pulmonary atresia (all forms)
Pulmonary hypertension complicating CHD
Pulmonary valve regurgitation (moderate to severe)
Pulmonary valve stenosis (moderate to severe)
Pulmonary vascular obstructive disease
Sinus of Valsalva fistula/aneurysm
Subvalvar or supravalvar aortic stenosis
Tetralogy of Fallot
Total anomalous pulmonary venous connection
Transposition of the great arteries
Tricuspid atresia
Valved conduits
Vascular rings
Ventricular septal defects with:

- Aortic regurgitation
- Aortic coarctation
- History of endocarditis
- Mitral valve disease
- Right ventricular outflow tract obstruction
- Straddling tricuspid and/or mitral valve
- Subaortic stenosis

Modified from Therrien J. et al. CCS Consensus Conference 2001 update: recommendations for the management of adults with congenital heart disease Can J Card 2001;17:940-59, 1029-1050, and 1135-1158 (with permission).

- Physicians without specific training and expertise in adult CHD should only not manage adults with moderate and complex CHD independently but in collaboration with colleagues with advanced training and experience in the care of such patients (usually based in a regional adult CHD centre).
- Patients with moderate or complex CHD may require admission or transfer to a regional adult CHD centre for urgent or acute care.
- Most cardiac catheterisation and electrophysiologic procedures for adults with moderate and complex CHD should be performed at the regional adult CHD centre, where appropriate personnel and equipment are available. If such procedures are planned at the local cardiac centre, prior consultation with adult CHD cardiology colleagues should be sought to avoid unnecessary duplication of invasive procedures.
- Cardiovascular surgical procedures in adults with moderate and complex CHD should generally be performed in a regional adult CHD centre with specific experience in the surgical care of these patients[9].
- Appropriate links should be made for provision of non-cardiac surgery. The need for developing an integrated team of high-risk obstetricians, anaesthetists and adult CHD cardiologists cannot be overstated.
- Each regional centre should develop a joint medical and surgical database, to record activity and outcomes, audit results and facilitate research. Comprehensive patient clinical records should be kept in the regional adult CHD centre and copied to the primary care physician and the individual patient.

Manpower, training and research

The importance of adult CHD as a sub-specialty of cardiology has been recognized by the Calman UK training advisory committee and the 2001 Bethesda Conference. Basic training in adult CHD is now mandatory for adult cardiology trainees. It is also recognized that selected individuals will need to train more comprehensively in the field. The American College of Cardiology Task Force states that a minimum of two years of full-time CHD training is needed for a senior cardiology trainee (with an adult or paediatric cardiology background) to become clinically competent, to contribute academically, and to effectively train others[11]. The small number of available centres which can offer comprehensive training in CHD at present, coupled with a shortage of resources remain major obstacles in achieving this goal. Training programs for other key staff (e.g. nurses, obstetricians, imaging staff, technicians, psychologists) in adult CHD teams should also be established. The first set of guidelines for the management of the adult with CHD, commissioned by the Canadian Cardiovascular Society, was recently revised by an international panel of experts[10] and is now available on the Internet (www.rbh. nthames.nhs.uk/cardiology/consensus). These guidelines have been endorsed by North American, European and other professional bodies. National and international curricula in adult CHD are being developed to disseminate existing information on the management of the adult with CHD and stimulate research. New communications tools such as videoconferencing and the electronic mail are bound to facilitate this process further.

Educational material to guide adult CHD patients is being developed. Advice on employability, insurance, pregnancy and contraception, exercise, endocarditis prophylaxis and non-cardiac surgery is being made available. Barriers to multi-disciplinary services should be challenged with the objective of making expert resources available for all adult patients with CHD who need them.

A new group of specialized cardiologists in adult CHD is required to ensure the delivery of high quality life-long care for this patient population which has benefited so much from early pediatric cardiology and cardiac surgery resources and expertise. Furthermore, there is a pressing need for clinical research on potential factors influencing the late outcome of this expanding patient population. The effects of medical, catheter and surgical intervention need to undergo further prospective assessment. Clinical and research resources must, therefore, be secured for this large patient population.

Transition of care

Structural plans for transition from pediatric to adult CHD care are being developed. Different models are applied depending on local circumstances. Individual patient education regarding their diagnosis and specific health behaviors should be part of this process. Comprehensive information including diagnosis, previous surgical and or catheter interven-

tions, medical therapy, investigations, current outpatient clinic reports and medication should be kept by the patient and also be sent to the adult CHD facility. The development of a patient electronic health "passport" is to be encouraged and is of particular relevance to patients with complex diagnoses and numerous previous interventions.

There is an international consensus that the multiple needs of this population can be best fulfilled through national frameworks with the following objectives:

- to establish a Network of Regional Centres for the adult with CHD
- to foster professional Specialist Training in adult CHD
- to co-ordinate National or Local Registries for adults with CHD
- and to facilitate research in adult CHD

Such a model of care, training and research for the adult with CHD would be in keeping with the 2001 Bethesda Conference and recent UK National Health System guidelines and has been implemented for some time in Canada. Within this framework, general cardiologists with an interest need to be supported locally in district general hospitals and be facilitated to work with both tertiary and primary care physicians to provide for the adult with CHD. Paediatric cardiology expertise must be utilized and transition care programs developed to ensure seamless care for this patient population. Patients, and their families, need to realize that life-long follow-up is required for most of them and that they may well require further intervention, medical and/or surgical, preferably before overt symptoms develop. Databases shared amongst paediatric, adult and non-tertiary care centres and easy access to regional facilities should be in place to promote this multi-level collaboration. Patient advocacy groups need to continue to develop and participate actively in this dynamic process.

In summary, adults with CHD are not any longer a rare or "odd" patient group. Many or most need expert life-long cardiac care. The time has come for a National Adult Congenital Heart Service[12], supported by the Departments of Health, relevant professional bodies and funding agencies, to care for the beneficiaries of this astonishing success story in the management of congenital heart disease.

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