Grown-up congenital heart disease - experience in a district general hospital

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Abstract

Some 340 adult patients (186 male, 154 female; average age 36 years) with congenital heart disease are now seen in a dedicated clinic at a district general hospital. Septal defects and aortic pathology account for 48% of cases seen and 21% have complex congenital heart disease. A first operation has been performed in 55%, a second operation in 13.3% and a third operation in 3.2%. Pulmonary hypertension is present in 7%. Eighty two of the 154 women have had 123 pregnancies. Care issues relating to the pregnant grown-up congenital heart disease (GUCH) patient are discussed. The growth of this population is highlighted, as is the requirement for more structured care. Issues relating to the establishment of a dedicated GUCH clinic are discussed, including training of cardiologists in this sub-speciality.

Key words: grown-up congenital heart disease in a DGH; setting up a GUCH clinic in a DGH; pregnancy care issues of GUCH in a DGH.

Introduction

The incidence of congenital heart disease is seven to 12 per 1,000 live births; 80% of these cases, including complex conditions, reach adulthood. Hunter has estimated that there may be more than 60 000 patients over the age of 16 years in the UK who need medical or surgical management of their congenital heart disease.[1] In the US it has been estimated that the figure would reach 900 000 survivors of congenital heart disease in 2000.[2] Figures from the Society of Cardiothoracic Surgeons of Great Britain and Ireland suggest 3–4,000 operations are performed on children with congenital heart disease each year, with very low surgical mortality.

Adults with congenital heart disease are still a relatively small population and it is acknowledged that more structured management for this population is required. There are a number of problems for this group: first, in the early days of cardiac surgery the operative intervention was considered to be a total correction and many patients were discharged and lost to follow-up; second, at the age of 16, patients can be discharged from the paediatric service without formal arrangements for ongoing care in adult cardiology, a problem that has recently been highlighted by Gatzoulis *et al*;[3] third, even if further follow-up is arranged, it may be that such patients are seen routinely on the junior staff list, who are unaware of specific complications or possible new interventions. Moreover, when such patients are scattered like rare flowers through many different clinics, consistency of care is very difficult. The temptation to keep 'interesting' clinical signs in a general clinic needs to be resisted.

Tertiary GUCH centres in the UK are vital for the management of complex congenital heart disease. They have the core expertise and also surgeons skilled in the problems of GUCH patients. Regional centres should be established to provide specialist care for the two main groups: those with more simple congenital heart disease who can usually be looked after without referral to a tertiary centre; and those with more complex congenital heart disease who can be seen on a 'shared care' basis.

Against this background a dedicated clinic for adults with congenital heart disease was started in 1993 in a Norfolk district general hospital. Initially the clinic was held once a month: it became twice-monthly in 1995, weekly in 1999 and its frequency continues to expand.

Patient population

The clinic sees 340 patients with an average age of 36 (range 16–71 years). Table 1 summarises the data.

Operative intervention has been performed in 186 patients (55%), with 45 (13.3%) requiring a second operation, 11 (3.2%) requiring a third operation and one requiring a fourth operation. Two patients have had a cardiac transplant. Recent non-surgical interventions have included four atrial pathways (Mustard operation) dilated and stented; two aortopulmonary (AP) collaterals dilated and stented; and umbrella occlusions in three patients – one residual duct and two closures of residual Potts anastomoses.

Pulmonary hypertension is present in 24 (7%); 21 are cyanotic and three are acyanotic. Seven patients have trisomy 21 (all with septal/atrioventricular [AV] canal defects). Other diagnoses are ventricular septal defect (VSD) in five, primum atrial septal defect (ASD)/AV canal in six, complex pulmonary atresia (CPA) in three, patent ductus arteriosus (PDA) in one, total anomalous pulmonary venous drainage (TAPVD) in one, and secundum atrial septal defect (ASD) in one. Venesection – with concomitant

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Table 1. Summary of GUCH patient diagnoses in a district general hospital

	Total number of patients	Male	Female
	340	**186**	**154**
Septal defects (28.2%)	**96**		
VSD	44	28	16
1°ASD/partial AV canal	2	16	15
2°ASD*	31	10	21
Aortic pathology (27.7%)	**94**		
Bicuspid aortic valve*	38	25	13
SubAS	13	7	6
SupraAS	2	2	
AR 2° con abn valve	2	1	1
Coarctation of aorta	39	26	13
Marfan/HOCM(15%)	**51**		
Marfan's syndrome*	41	29	12
HOCM*	10	5	5
Fallot's tetralogy (10.6%)	**36**	**19**	**17**
Other congenital valvular abnormalities (7.9%)	**27**		
Pulmonary stenosis	17	8	9
Mitral valve lesions	9	2	7
Ebstein's anomaly	1		1
Transposition types (5.3%)	**18**		
TGA	15	10	5
CCTGA	3	3	
Complex congenital heart disease (5.3%)	**18**		
Single ventricle	9	2	7
Complex pul atresia	5	2	3
Pulmonary atresia/intact VS	2	1	1
Tricuspid atresia	2		2

Key: * patients also seen in general cardiology clinics
1°ASD = primum atrial septal defect; 2°ASD = secundum ASD; AR = aortic regurgitation; AV = atrioventricular; CCTGA = congenitally corrected transposition of great arteries; complex pul atresia = complex pulmonary atresia with multiple aorto-pulmonary collaterals; con abn = congenital abnormality; HOCM = hypertrophic cardiomyopathy; subAS = subaortic stenosis; supraAS = supra-aortic stenosis; TGA = transposition of great arteries; VS = ventricular septum; VSD = ventricular septal defect;

fluid replacement – is currently required in nine patients. Of the three acyanotic patients, the underlying diagnosis is repaired double outlet right ventricle (DORV) in one, repaired PDA/VSD in one and repaired PDA and coarctation in the last.

82 of the 154 women have had 123 pregnancies. Six of the resulting 123 children (5%) have congenital heart disease – this figure does not include those families with hypertrophic cardiomyopathy (HOCM) or Marfan's syndrome. There have been 10 mid-term miscarriages. The underlying diagnoses in these women include 11 tetralogy of Fallot, 11 Marfan's syndrome, nine aortic valve disease, six pulmonary stenosis, six coarctation of the aorta, eight ventricular septal defect, six secundum ASD and four primum ASD; five subaortic stenosis, five HOCM, five mitral valve lesions; four transposition of the great arteries (TGA), one DORV, and one Ebstein's anomaly. Five out of 154 women have been sterilised because of a contraindication to pregnancy and four patients have been advised against pregnancy because of pulmonary hypertension.

Since 1993, there have been 10 deaths in patients attending this clinic (this figure is not included in the 340 patients currently on the database). One was catheter-related TGA/DORV at a tertiary centre), one was perioperative (a third operation for VSD/AR), three were related to severe heart failure awaiting transplantation (CPA, tricuspid atresia [TA], pulmonary hypertension and cor triatriatum), one endocarditis with cerebral abcess (TGA/VSD/Blalock-Taussig shunt/PA band), two with ventricular tachycardia/ventricular fibrillation (both with AV canal and poor left ventricular function), one sudden death at night (TGA), one pregnancy-related (closed secondum ASD, pulmonary artery pressure of 50 mmHg; pregnancy supervised at tertiary centre because of the high risk stated to the patient pre-conception).

Septal defects

Septal defects are the largest group in this GUCH population. Four of these patients have trisomy 21 and have not been repaired; all have pulmonary hypertension. The remaining patients are continuing follow-up because of impaired right or left ventricular function, rhythm disturbances or other residual lesions, including aortic regurgitation (AR) in five patients and pulmonary hypertension in a further five.

Fifteen out of 21 patients with a primum ASD/partial AV canal have had this repaired. Pulmonary hypertension is present in six patients with open defects, of whom three have trisomy 21. Follow-up of patients with secondum ASD is ongoing because of secondary clinical problems, including arrhythmias, poor right ventricular function, pulmonary hypertension in one patient and pacemaker implantation in another.

Aortic pathology

Patients with aortic pathology are the second largest group, of whom 17 have undergone operations – seven with a previous valvotomy and four with reoperations. Patients with isolated bicuspid valves are seen in general clinics as well. Of the 13 patients with subaortic stenosis, 11 have been operated upon.

Coarctation repair was as follows: end-to-end anastomosis in 23 (five reoperated); subclavian flap angioplasty in six (one reoperated); five patch angioplasty (one reoperated); four graft repairs; and one native coarctation referred for surgery. The associated lesions in this group are bicuspid aortic valve in 19 (33%), monocusp aortic valve with left ventricular descending aorta Hancock valve in one, AR in five and Turner's syndrome in two. Twelve (21%) are hypertensive and all non-hypertensive patients are screened annually with ambulatory blood pressure monitoring.

Marfan's syndrome and hypertrophic cardiomyopathy

Of the patients with Marfan's syndrome 10 have now been operated upon; 26 have a positive family history. Two patients with

HOCM have had a myomectomy and one has a DDD pacemaker; five patients have a positive family history.

Younger patients with these diagnoses are seen in this clinic but also other general cardiology clinics. (A further 75 patients with HOCM have been identified as receiving follow-up in general clinics.)

Fallot's tetralogy

In this patient group, 14 have had a second operation, four have had a third operation, while one patient (aged 71 years) has never had this operated on and may be one of the oldest non-operated survivors. A right aortic arch is present in 15%; other associated lesions include atrial septal defects in three, persistent left superior vena cava in two and absent left pulmonary artery in one. Significant pulmonary regurgitation (PR) is present in 12, residual right ventricular outflow tract obstruction (RVOTO) in five, residual VSD in seven, poor right ventricular function (in the absence of PR) in four and poor left ventricular function in two. Aneurysms of the RVOT have been detected in two patients and operated on in one. Rhythm disturbances are a problem in nine patients – atrial flutter in four and supraventricular tachycardia in five; three patients have pacemakers.

Other congenital valvular abnormalities

Three patients with pulmonary stenosis are deaf and one has mental retardation. Of the patients with mitral valve lesions (which include parachute and cleft mitral valves), four have been operated on.

Transposition of the great arteries

Atrial repair of this condition has occurred in 14 (11 Mustard and three Senning); one patient has had a Rastelli repair. Four patients have pacemakers and atrial flutter is a problem in three. Baffle pathway stenoses have been dilated and stented in four patients. One patient has a baffle leak associated with mild desaturation with exercise but venous access is a problem for any intervention.

Two patients with congenitally corrected transposition of the great arteries have pacemakers, of whom one is currently being investigated with a view to tricuspid valve replacement.

Complex congenital heart disease

The single ventricle is DORV in four patients, double outlet left ventricle (DOLV) in one patient, double inlet left ventricle (DILV) in one patient and indeterminate in three patients. Associated lesions include single atrium in two patients, TGA in two patients, dextrocardia in one patient and coarctation in one patient. Operative intervention has been performed in seven patients, with septation in five, total caval pulmonary connection (TCPC) in one, Blalock-Taussig and Glenn shunts in one.

Of the patients with CPA, three have shunts, one has a right ventricular-pulmonary artery (RV-PA) conduit and one is unoperated with multiple aorto-pulmonary collateral arteries (MAPCAs) (no stenoses). One patient with pulmonary atresia with an intact ventricular septum has a shunt and the other has both a shunt and a conduit. Both patients with tricuspid atresia have had the Fontan operation. Within this group are two patients seen in this GUCH clinic for the first time with unoperated complex congenital heart disease; one is well six years after a TCPC for single ventricle/single atrium (SV/SA) and the other is well five years after a series of staged operations leading to an RV-PA conduit for CPA with ventricular septal defect.

Discussion

Patient population

The distribution of diagnoses described above is probably fairly typical of the sort of patients it might be expected to see in a district general hospital, with the majority being simple congenital heart disease. Wren and O'Sullivan have recently suggested that an adult population with congenital heart disease would comprise 28% complex, 54% significant (requiring intervention) and 18% minor cases (eg. ventricular septal defect, aortic stenosis, pulmonary stenosis not requiring intervention at one year). A broad comparison with this clinic yields 7.3% complex, 65% significant and 27.7% minor cases. These figures exclude the patients with Marfan's syndrome and cardiomyopathy seen in this clinic, which Wren and O'Sullivan have highlighted as requiring follow-up.[4]

Nearly 60% of the female patients described have had a pregnancy, and care of such patients is clearly important in a district general hospital clinic. Ideally, every woman with congenital heart disease should be assessed prior to conception to ensure an accurate diagnosis of her underlying condition. Patients with pulmonary hypertension and Eisenmenger's syndrome are at such high risk that everything should be done to terminate or prevent pregnancy. It is important to assess the haemodynamic consequences of the pregnancy on the cardiac disorder and, in turn, of the cardiac disorder on the baby's development.

The pre-conception visit is also a good time to discuss the risks of congenital malformation in the offspring. Recurrence risks can be precisely defined when a syndrome is caused by mutations at different sites of a single gene.[5] A visiting clinical geneticist allows full discussion of inheritance if indicated. We maintain close liaison with obstetric, fetal medicine and anaesthetist colleagues and see patients routinely at 12–14 weeks and again in the third trimester – although more frequently if problems are predicted or occur.

More complex congenital heart conditions can be managed in consultation with a tertiary centre. It is reasonable for them to be managed at their district general hospital as long as potential problems are discussed early with the tertiary centre. Furthermore, patients often find travelling to a tertiary centre expensive and time-consuming. There is a rationale which suggests shared care may be preferable in many cases, for when patients are acutely unwell or when they develop a non-cardiac condition they will be seen at their local district general hospital, which will then have the cardiac condition documented in the notes.

The GUCH clinic

We have some general comments about running a GUCH clinic in a district general hospital, which are highlighted in table 2.

Table 2. GUCH clinics in a district general hospital – relevant topics

- Adequate documentation of previous operations/interventions and catheters is necessary. Obtaining the original operation report is sometimes difficult but is vitally important because problems and anomalies present at the operation are revealed which are often pertinent to the current position
- Longer clinic slots are needed due to the complicated medical problems; GUCH patients often need advice on contraception, pregnancy, adoption, risks to offspring, fitness to drive or fly, housing, life insurance, social service support (including disability living allowance), exercise, dangerous hobbies and psychological problems
- Highlighting the existence of the GUCH clinic locally through regular talks to junior staff (who change frequently) and local GPs, emphasising the need for follow-up
- Liaison with tertiary centre needs the availability of discussion of cases by telephone or e-mail. Junior staff should ensure that copies of letters to GPs are also sent to the district general hospital cardiologist. There is a role for videoconferencing
- Local investigations – ECG, Holter and ambulatory blood pressure monitoring, exercise testing +- finger pulse oximetry, transthoracic and transoesophageal echocardiography, magnetic resonance imaging, nuclear cardiology, ambulatory monitoring, catheterisation, pacing
- Liaison out-patient facilitator (clinical nurse specialist/secretary/ receptionist) to provide a telephone helpline to give support, advice or arrange an urgent out-patient appointment
- Role of national database – size of the GUCH patient population is unknown
- Secretarial support – more letters generated/patient in terms of support for life assurance, help with work applications/DHSS details
- Sub-specialist year for those in training

Table 3. Issues to be highlighted at first clinic visit

- Patient's understanding of his condition – 'a well informed patient can potentially act as an avenue of communication between health care providers'[6]
- Introduction to GUCH patient network (Helpline No: 0800 854 759; www.guch@demon.co.uk; 12 Rectory Road, Stanford-le-Hope, Essex, SS17 0DL)
- Endocarditis protection (new card if necessary)
- Issues relating to contraception and pregnancy
- Lifestyle issues – tobacco, alcohol, drugs, exercise
- Lifestyle issues – jobs, insurance etc.
- Lifestyle issues – psychosocial
- Introduction to liaison nurse as a resource

Clinic slots do need to be longer than usual since these patients not only have complicated medical problems requiring attention, but frequently need advice about other aspects of their lives. There is a tendency for paediatric cardiologists who have been seeing patients biannually to suggest that the follow-up in adult service is arranged for two years hence. We feel that these patients need to be seen sooner – perhaps even after six months for the first visit. This is because these adolescents are often leaving school for jobs, college or university and it is a time of turmoil for them. Appointments two years hence with a new face may not be very appealing, or even remembered.

Irrespective of whether the patient has been referred on from the paediatric service or from another clinic, we take the opportunity to examine a number of topics, highlighted in table 3. It is important to assess what the patient understands about his diagnosis and operations. This is especially pertinent if the patient has always been accompanied by a parent who knows the answers; nevertheless it is relevant in all age-groups.[6] Advice about endocarditis pertains not only to dentistry but also to tattooing[7] and body piercing.

The liaison out-patient nurse provides a telephone helpline. At this hospital she is a clinical nurse specialist (CNS) so titration of prescribed medication, arranging haematology/biochemistry investigations, organising electrocardiograms, echocardiograms, Holter monitoring, and so on, are all part of her remit; she may also arrange earlier consultant review or admission as required.

The CNS is also involved in arranging venesections for the GUCH patients with polycythemia in an elective day case admission bay. Important considerations in venesecting these patients include; limiting venesection to 250 ml, administering concomitant plasma expanders, and using filters with the IV giving sets to protect against small air bubbles.

We have identified a link cardiology ward where GUCH patients can be admitted directly under the cardiology team; the ward sister and nursing team have taken a lead in increasing their knowledge of congenital heart disease. We have particular concerns about these patients being reviewed in accident and emergency or being admitted acutely for surgery or with new arrhythmias without the admitting doctor realising the significance of their congenital heart disease. As a result of a newly installed computer system at this hospital, we have flagged up 'alerts' on these patients so that early discussion can take place.

Communication between centres

Shared care of patients with congenital heart disease engenders learning, co-operation and discourse between cardiologists at district general hospital level and at tertiary centres. Tertiary centres frequently have joint clinical-pathological conference (CPC) meetings where cases are presented and discussions occur between interventionists and surgeons as to the best approach to treatment. Although presenting patients at these meetings for the district general hospital cardiologist is informative and helpful, it is not practical on a regular basis because of time and distance constraints. Ideally it should be possible to set up a system to allow videoconferences between the tertiary centre and any number of district general hospitals on a regular basis.

Facilities and resources

Congenital heart disease in the adult and the associated surgical corrections are technically challenging for the echocardiographer. It is helpful to have the full operation report at the time of carrying out echocardiography on these patients to allow assess-

Key messages

- The requirement for grown-up congenital heart (GUCH) clinics is predicted to increase markedly as more survivors reach adulthood. Key issues relating to such clinics are reviewed
- Specific GUCH clinics allow recognition of known diagnosis-related complications and possible interventions which may not be considered in general clinics.
- 'Shared care' engenders learning, co-operation and discourse between district general hospitals and tertiary centres. The cardiac condition will also be documented in notes in case of emergency non-cardiac admissions locally
- Transition from the paediatric to adult service is an important time to emphasise issues about diagnosis, endocarditis risk, lifestyle/job/psychological issues and the GUCH patient association
- Pregnancy in these patients is increasing markedly and requires planned shared care with the obstetrician and obstetric anaesthetist

ment of the original anatomy and how this has been changed by the surgery. Patients with complex disease and/or surgical correction cannot be imaged adequately in 10 minutes. At the first appointment, patients with complex pathology tend to be echoed jointly with the cardiologist; the anatomy is drawn onto the report form. In addition to the standard views, we have found that imaging from the high parasternal and right parasternal views will often allow us to see pathways that cannot be seen in the standard views. Transoesophageal echocardiography has also enhanced our ability to image these patients, particularly in delineating baffle pathways (in TGA), vegetations, thrombus, atrial septal defects and pulmonary venous drainage.

We are fortunate in having magnetic resonance imaging (MRI) scanning (with cardiac software) to complement non-invasive investigations. Nuclear cardiology, particularly to obtain assessment of right ventricular ejection fraction with first pass techniques, is also useful. We have cardiac catheterisation available at this district general hospital but, in some instances, this may be best performed at the tertiary centre, particularly if a further intervention that may be completed at the same time is a possibility.

A number of GUCH patients need pacemakers. When they do, we believe it essential that they are paced physiologically (ie. with atrial or dual chamber systems) provided that they are not in permanent atrial fibrillation. A cardiologist who is experienced in physiological pacing and the use of screw-in electrodes will rarely encounter insurmountable problems with endocardial pacing; indeed, these patients provide a rewarding technical challenge. In some cases venous anatomy needs to be delineated first.

Training

District general hospital cardiologists seeing adult patients with congenital heart disease will no doubt need to demonstrate continuing specific GUCH medical education, in the future. Adult cardiologists in training will need to spend one year in a tertiary centre to understand the anatomical substrate and specific condition-related complications. Paediatric cardiologists in training will similarly need to spend one year in adult cardiology to understand some of the non-congenital issues that will arise in these patients. There needs to be further agreement concerning GUCH training as a subspecialist option in year six of a specialist cardiology registrar's training.

Conclusions

Adults with congenital heart disease are a growing medical community who need special care and attention. As they are still a relatively uncommon group, it is difficult for those who see them rarely to appreciate the dangers which such patients face. It is important that we do not allow these patients to be lost to follow-up. One in five deaths of such patients is said to be avoidable or premature.[8] It has also been pointed out that they are an expensive group of patients, both in terms of resources and time.[9] Provision will need to be made for this. Gatzoulis and colleagues reported a 265% expansion in patient workload over 10 years in Toronto[3] and Wren and O'Sullivan a 400% increase in the number of clinics for adults with congenital heart disease over the same time period in Newcastle.[4]

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