Current perspectives

Life expectancy and quality of life in adult patients with congenital heart disease

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The survival and quality of life of patients with congenital heart disease have significantly improved in the last 20 years. This is due to more effective medical and surgical care. The new community of grown-up congenital heart patients consists of a few natural survivors with trivial congenital lesions or very rare complex cardiac abnormalities which are naturally compensated, and of more than 75% of patients who had been submitted to cardiac surgery during infancy or childhood. Clinical follow-up is however mandatory for many of them with scheduled times and types of exams to control the effects of sequelae and late complications, and to prevent deterioration and premature death because cardiac surgery may not have resulted in normality.

Moreover, these patients have many needs and even more, many questions. Not giving a correct answer to each specific question reduces the entity of surgical success.

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In the absence of surgery, few congenital heart defects are compatible with a disease-free survival to adult life. Since the middle of the 1970s, the success of cardiac medical and surgical care has dramatically increased the number of newborns who have survived through infancy, reached adolescence and became adults, constituting a new medical community referred to as GUCH (grown-up congenital heart disease) patients. In the last 15 years we have observed a significant rise not only in the size of this population but also in the proportion of patients with complex lesions. If their quality of life is to be satisfactory, specialized medical care and particular attention are necessary[1,2].

Grown-up congenital heart disease population

Unoperated patients without pulmonary vascular disease. At the present time, only 20-25% of the adolescent and adult patients admitted to the GUCH unit had not been submitted to cardiac surgery during infancy and/or childhood[3]. The majority of unoperated patients have a simple congenital heart defect such as an atrial septal defect or a small restrictive ventricular septal defect, or mild pulmonary stenosis or a bicuspid aortic valve defect[4]. Indeed, survival to adulthood is the rule for patients with an ostium secundum atrial septal defect, one of the most common congenital cardiac abnormalities. However, in the absence of surgery, their life expectancy is not the same as that of the general population because mortality after age 40 increases to approximately 6% annually. The late survival of patients undergoing surgery before the age of 24 years is the same as that of the age/sex matched control population. However, it is poor for those undergoing closure of atrial septal defects after age 41 due to an increased incidence of supraventricular arrhythmias: it is well known that atrial fibrillation carries the increased risk of cerebrovascular accidents[5,6].

Individuals with small ventricular septal defects, a normal heart size and a normal pulmonary vascular resistance are not at increased risk of death or disability[7]. Ventricular septal defects are the most common congenital heart disease associated with Eisenmenger's reaction in adulthood[8].

Ninety percent of untreated patients with complicated coarctation of the aorta (coarctation of the aorta associated with other intracardiac or extracardiac anomalies such as obstruction of the outlet of the left ventricle, ventricular septal defects, mitral valve abnormalities, intracranial aneurysms, Turn-

er's syndrome) die within the first year of life. Fifty percent of patients with simple coarctation (no other relevant lesions) who survive after the second year of life die before age 32, 75% before age 46, and 92% before age 60[9].

Patients with complex congenital cardiac anomalies who have not had surgical correction have a long survival only when favorable anatomical conditions permit a satisfactory pulmonary flow, without severe pulmonary hypertension, and a long preservation of ventricular performance (univentricular heart with pulmonary stenosis or complete transposition with ventricular septal defect and pulmonary stenosis, Ebstein's anomaly).

Unoperated patients with Eisenmenger's reaction. Seventy percent of the adult population with cyanotic congenital heart disease has pulmonary vascular disease[10]. The other cyanotic patients have untreated complex defects with severe pulmonary obstruction and collateral systemic arterial vessels with proximal stenosis or insufficient palliative aortic-pulmonary shunts: in these patients hematologic problems such as pulmonary or cerebral thromboembolisms or hemorrhagic events are common and play a significant negative role in their clinical course, thus determining the prognosis and quality of life[11]. Despite the severity of disease, many patients with pulmonary vascular disease (Eisenmenger's reaction) maintain a satisfactory quality of life and well being for a long time (80% of patients with an ability index of 1 or 2 at the first visit at the GUCH unit after age 9). Clinical deterioration in terms of a reduced effort tolerance or increased cyanosis and the onset of symptoms related to blood viscosity involve a large percentage of patients at a mean age of 25 to 29 years. Only 5% of patients have objective signs of congestive heart failure which develops later on, after a mean age of 42 years. Patients with complex congenital heart disease and Eisenmenger's reaction are subject to earlier relevant disabilities and clinical deterioration. Pulmonary and cerebral complications remain the most important clinical events which cause a deterioration in the quality of life and even death. Hemoptysis (21%) may be due to pulmonary infarction consequent to thrombosis *in situ* or when catastrophic, to vessel rupture; it may occur after a period of intense stress or excitement. Cerebral complications (stroke 8% and cerebral abscess 4%) profoundly influence the quality of life but do not significantly modify the survival: a hemoglobin plasma level > 18 g/dl is not associated with a significantly increased hazard ratio of cerebral complications[10]. Conventional wisdom assumes that a high hematocrit in cyanotic heart disease increases the risk of thrombotic incidents and its reduction by phlebotomy is therapeutically useful[12]. This is relatively true for polycytemia rubra vera, in which vascular occlusive events are common and most often take the form of cerebral arterial thromboses, with a strong positive correlation between the hematocrit and vascular occlusive episodes. However, during the erythrocytotic phase, even in these cases phlebotomy alone is associated with a statistically significant increase in the risk of thrombotic complications[13]. A risk which increases with frequent phlebotomy. In patients with congenital heart disease, hypoxia does not result in panmyelosis, such as polycytemia rubra vera, but only in an increase in the red cell mass with normal leukocyte counts and a platelet count generally in the lower range of normal when not significantly reduced. The red cell mean volume is increased in cyanotic heart disease. On the contrary, the red cells of patients with iron deficiency anemia are microcytes with a less pliable membrane which increase the blood viscosity despite the presence of a lower hemoglobin concentration[14]. Several patients with cyanotic congenital heart disease and low serum ferritin concentrations have a tendency towards elevated erythropoietin titers, suggestive of relative anemia. Symptoms of iron deficiency can mimic those of hyperviscosity but true symptomatic hyperviscosity rarely occurs in iron-replete cyanotic patients with hematocrits < 65%[15]. In such circumstances phlebotomy aggravates, rather than alleviates, the symptoms, moreover increasing the risk of cerebrovascular incidents. The right ventricular function, the presence of complex congenital heart disease and the age of onset of clinical deterioration are the variables which adversely affected the prognosis at uni- and multivariate analyses performed in a large European collaborative study[10]. On the basis of the suggested pathophysiological mechanisms that underlie the development of pulmonary vascular disease, the introduction of vasodilators[16], particularly high-dose nifedipine[17], in an attempt to reduce the pulmonary vasoconstriction which would accelerate the obstructive changes is justified. However, these patients differ considerably in their response to vasodilator agents with significant changes over time. In fact, their effects are exerted more on the systemic circulation and a paradoxical clinical picture including systemic hypotension and increased cyanosis and only a minimally reduced pulmonary vascular resistance may ensue. Finally, improvement after acute treatment may not imply long-term efficacy. The positive effect of long-term oxygen treatment[18] at home for a minimum of 12 hours, preferably 15, significantly improves the ability index rating in adult patients but does not modify survival. Recently, it has been found that long-term continuous intravenous prostacyclin therapy[19] administered with the aim of reducing the pulmonary vascular resistance has been more effective than calcium-channel blockers. In patients with primary pulmonary hypertension, the former therapeutic regimen exerted vasodilatory, antiproliferative and platelet-inhibiting effects. No clinical trial has used prostacyclins in obstructive pulmonary vascular disease secondary to congenital heart disease. A clinically applicable formulation for the administration of prostacyclins by means of

aerosol solutions and chemically stable oral analogs has yet to be developed[20]. Sporadic cases of patients with Eisenmenger's syndrome treated with sildenafil[21] have recently been reported, but the positive effects have to be confirmed.

Nevertheless, we can still significantly influence the natural history of the disease and the quality of life of patients with pulmonary vascular disease, by means of a correct management directed at the avoidance of the complications associated with pulmonary hypertension, chronic aortic desaturation, abnormalities of hemostasis, ventricular dysfunction, infections, dehydration, sudden vasodilation, drug side-effects, pregnancy, hormonal contraception, extracardiac surgery, and physical activity[22].

Patients operated upon for total correction. Certainly cardiac surgery has dramatically changed the natural history of simple and complex congenital heart diseases. The perioperative mortality has significantly improved in the last 20 years but the delayed outcome is not so brilliant.

If early surgical results are easy to quantify because of two alternative conditions, death or survival, the definition of late results of surgical correction becomes difficult by various terms such as native heart disease, anatomic modifications produced by surgery, relationship between the psycho-physical status of the patient and the reality in which he lives and has to compare, and institutions, concerning their capacity to satisfy driven needs. For these patients the substantiality between quality of health and quality of life is even more obvious. Owing to the fact that cardiac surgery may not necessarily result in normality, a thorough clinical follow-up with scheduled times and types of exams is the best way to accurately evaluate the effects of sequelae and complications and to prevent deterioration and premature death. An increased survival does not necessarily imply an improved quality of life. Thus, during postsurgical follow-up the percentage of patients free from significant pathological events has to be taken into consideration. Only 5% of patients operated upon for atrial septal defects, anomalous drainage of one pulmonary vein or pulmonary valve stenosis will develop delayed major clinical problems during follow-up. These complications arise in 10-15% of patients operated upon for ostium primum or subvalvular aortic stenosis, in more than 50% of patients operated upon for atrioventricular septal defect with a common valve, and in 100% of patients submitted to aortic valvulotomy, the Mustard or Senning operation or Fontan procedure[3].

The cumulative overall 30-year survival rate of patients with larger ventricular septal defects who have undergone surgical closure is 82% compared with 97% in age/sex-matched controls whose risk of sudden death ranges from 0.09 to 0.24% per year[7,23].

Coarctation repair by surgery or by percutaneous balloon angioplasty[24] does not normalize the survival rate, because of the persistence of arterial hypertension and owing to the onset of other cardiovascular complications (survival rate: 91% after 10 years of postsurgical follow-up, 84% after 20 years and 72% after 30 years). During effort an abnormal systemic blood pressure response can occur in patients with optimal repair particularly if the decoarctation procedure was performed at an older age[25]. Postsurgical aneurysms of the aorta have been reported in about 5-9% of patients[26].

Mustard and Senning operations (atrial switch) for transposition of the great arteries. Since the end of the 1950s, the atrial switch operation has resolved the problem of survival of newborns with simple transposition of the great arteries. The perioperative mortality is very low (an overall survival of 65% after 25 years of follow-up; 85% survival for patients with simple transposition, and 45% survival for patients with complex transposition) and these patients maintain an acceptable work capacity for a long time[27]. However, late right ventricular dysfunction and life threatening arrhythmias[28-30] caused the abandonment of this operation that was definitively supplanted by the arterial switch operation[31] in the middle of 1980s. Presently, a large number of patients admitted to the GUCH unit for congestive heart failure have a history including an atrial switch operation performed 20-25 years ago: many of them need heart transplant[32]. Conversion to anatomic repair after adequate left ventricular retraining has been proven to be a successful alternative to cardiac transplant; however, older age is an important risk factor[33]. In their experience with left retraining, Poirier and Mee[34] did not report any survivor in whom atrial switching had been performed after the age of 15. The same results were obtained by other surgical teams with series not exceeding 5 patients[35,36]. The longer the morphological left ventricle works at a low pulmonary pressure, the less the capability of inducing left ventricular hypertrophy. A moderate hypertension of the subpulmonary ventricle secondary to pulmonary hypertension or to pulmonary or subpulmonary obstruction might play an important role in maintaining the capability of the myocardium to produce an adequate hypertrophic response after programmed pressure overload (banding). Indeed, 22 years after a Mustard procedure, we successfully performed[37] conversion of atrial to arterial switch repair prior to left ventricular retraining in a 23-year-old woman with severe right ventricular dysfunction. She had moderate pulmonary hypertension (58/21 mmHg, mean 40 mmHg) with normal pulmonary arteriolar resistance (1.5 U/m^2). The pulmonary banding was maintained for 8 months, with an adequate increase in left ventricular mass, which permitted a successful arterial switch conversion, improvement in the right ventricular pumping function and in the tricuspid valve insufficiency. After 3 years of follow-up, the patient shows an ability index of 1.

The incidence of delayed sudden death after a Mustard or Senning operation varies from 2 to 8% within a maximum of 10 to 15 years after surgery[38,39]. In his literature review including 104 cases of sudden death after a Mustard or Senning operation, Gilljam[40] reports that most sudden deaths appear to occur within 3 years of the operation. There is a second cluster of deaths occurring between 9 and 12 years after surgery. This latter datum is also supported by the Great Ormond Street Hospital data published by Gewillig et al.[30]. About half of the patients who died suddenly had current or previous hemodynamic abnormalities, atrioventricular blocks or atrial tachyarrhythmias but about one third had an apparently good hemodynamic status with no arrhythmias. Sixteen percent of the sudden deaths occurred in patients with asymptomatic sinus node dysfunction with bradycardia. Before surgery, all patients with transposition of the great arteries show a sinus rhythm. Seven years (mean interval) after atrial switching, only 50% of patients maintain this prerogative: junctional rhythm, atrial flutter and sinus node dysfunction are the most frequent arrhythmic abnormalities[41]. Atrial flutter seems to be a significant risk factor for sudden death. Moreover, atrial flutter is correlated with ventricular dysfunction. The mechanism of sudden death in patients with atrial flutter is speculative: supraventricular tachyarrhythmia associated with altered atrial contractility constitutes a significant impediment to ventricular diastolic filling and is a cause of a reduced systemic pressure and an insufficient coronary supply. In 36% of patients who had been submitted to atrial switching[42], ventricular tachycardia could be induced at electrophysiologic studies. The role of asymptomatic dysfunction of the sinus node associated with bradycardia has been discussed, but this condition seems to be an unimportant risk factor. An increased occurrence of junctional rhythm not associated with a mutual rise in the risk of sudden death during follow-up is well defined in these patients. Transvenous pacemaker insertion is required for symptomatic bradycardia or in case of the occurrence, at 24-hour Holter monitoring, of sinus node arrest with a pause of > 3.5-4 s. Antitachycardia pacing is indicated for some atrial arrhythmias. The insertion of a permanent pacemaker does not always prevent sudden death and 8% of patients with such a device died suddenly[43].

Anatomical correction (arterial switch). As far as the outcome following the arterial switch operation is concerned, we know that about 15 years after the first operations, left ventricular size, mass, functional status and contractility continue to be normal, with no evidence of a time-related deterioration in function[44,45].

Nevertheless, the rapid two-stage arterial switch operation constitutes a higher risk for mild impairment of myocardial mechanics. Serial evaluation indicates a slight but significant trend towards ventricular dilation, which is perhaps related to a relatively high incidence of at least mild aortic regurgitation (30%). Supravalvular pulmonary artery stenosis has been noted but it is almost always mild and usually does not show any tendency to progress. Supraventricular arrhythmias are very rare but exercise-induced ventricular tachycardias have been reported. The latter are probably related to local defects in myocardial perfusion[46].

Total correction of tetralogy of Fallot. The early results of surgical correction of tetralogy of Fallot are very good, < 1% of perioperative deaths. The actuarial 10, 20, 30 and 36-year survival rates of a large series of patients who survived through the first year after surgical repair[47] were 97, 94, 89 and 85%, respectively. The long-term results of surgical correction of tetralogy of Fallot in infancy and childhood are good in terms of health assessment and exercise capacity[48]: 89% of the long-term survivors described their health as "excellent" or "good", and 79% had an almost normal exercise capacity > 80% of the predicted value. A good health assessment is an important determinant for the quality of life, but we did not find any correlation between a "good" or "excellent" health assessment and the absence of symptoms (decreased exercise capacity) or sequelae (right ventricular dysfunction, ventricular or supraventricular arrhythmias). If the long-term results of surgical repair were to be judged on the basis of the presence of a normal cardiac anatomy or electrophysiologic findings, the score would not be good: as a consequence of pulmonary and tricuspid valve incompetence, 60% of patients had a substantial dilation of the right ventricle 5 years after surgery and 90% after 15 years; at delayed follow-up, 24-hour ECG revealed that 75% of patients had ventricular or supraventricular arrhythmias, with an incidence of sudden death of 0.3-0.4/1000 per year[49]. The factors related to sudden death may be mechanical, such as a high residual ventricular pressure, or anatomic such as increased fibrosis particularly in patients operated upon late (> 5 years of age). Moreover, fibrous-fatty substitution characterizes the area around the scar, a residual effect of ventriculotomy. All these lesions might provide substrates for focal abnormalities in ventricular depolarization and repolarization and can be utilized for the stratification of arrhythmic risk. In fact, clinical recordings of these alterations form the basis of the identification of those patients who are at risk for the development of ventricular arrhythmias related to sudden death. We found a statistically significant difference between the amplified ECG parameters of patients operated upon for tetralogy of Fallot with minor ventricular arrhythmias and those of patients with severe arrhythmias[50]. However, in these patients the presence of complete right bundle branch block reduces the clinical significance of the presence of delayed potentials to a different extent to that of the high accuracy demonstrated by this exam in patients with myocardial infarction or arrhythmogenic right ventricular cardiomyopathy. Owing to the corre-

lation between the QRS duration and the end-diastolic right ventricular volume, a QRS duration > 180 ms is considered by Gatzoulis et al.[51] as an important predictive parameter for ventricular tachycardia. On the basis of our data we believe that the clinical significance of the QRS duration is reduced by the presence of complete right bundle branch block, in view of the fact that we were unable to distinguish the increase in the QRS duration secondary to postsurgical damage of the conductive tissue from that consequent to right ventricular dilation. We first described the clinical significance of temporal and spatial QT dispersion as independent predictive factors for ventricular instability in patients operated upon for total correction of tetralogy of Fallot by ventriculotomy with a cut-off of 80 and 65 ms respectively. In our series of 66 patients[50], the mean age at operation was 11 years and the mean length of postsurgical follow-up was 16 years. QT dispersion and right and left ventricular function were the discriminating parameters between patients with and without ventricular tachycardia. On the basis of the values of QT dispersion and end-diastolic right ventricular volume, we can calculate the probability of sustained ventricular tachycardia in patients operated upon for total correction of tetralogy of Fallot. For example, with an end-diastolic right ventricular volume of 100 ml/m^2, according to the different values of spatial QT dispersion, the probability ranges from 0 to 30% with a pathological cut-off of 80 ms. Besides, in this adult population of patients right ventricular dysfunction appears to be the most important factor of the postsurgical history and determines electrical instability and late-onset progressive heart failure. Several recent reports correlate pulmonary insufficiency with right ventricular dilation and a reduced capacity for exercise[52]. In the presence of right ventricular outflow tract stenosis, pulmonary artery branch stenosis, residual ventricular septal defects and tricuspid incompetence, pulmonary incompetence is poorly tolerated. These data should be borne in mind when deciding the optimal timing for operation or re-operation and the nature of the corrective surgery[53,54]. The core of clinical follow-up is to avoid these disadvantages by means of appropriate surgery or of an interventional catheterization technique. Current surgical policy, aimed at the early and complete surgical correction, results in increased transannular patch rates with more residual pulmonary regurgitation and stenosis of the pulmonary arteries. The reasons leading to the optimism that modifications in the surgical technique and younger age at the time of repair might decrease the incidence of delayed sudden death in the last generation of patients operated upon via the transatrial approach should be reconsidered.

Fontan operation. The precariousness of the Fontan operation, initially introduced as a palliative therapy for patients with tricuspid atresia and then extended to various forms of univentricular circulation, is documented by the high number of variants suggested in order to limit major long-term complications which, in the late follow-up, reduce its efficacy: onset of supraventricular tachyarrhythmias, progressive deterioration in ventricular function and non-homogeneous distribution of the pulmonary vascularization[55]. The mean survival of these patients 10 years after surgery is 60% (range 80-51%, depending on the indication)[56]. The functional status 5 years after operation is one NYHA functional class higher than before the operation in 30-35% of patients, in the same class in 16-20%, and in a lower class in 35%[57]. Patients with optimal surgical results have a reduced work capacity[58,59].

To reduce the incidence of delayed supraventricular arrhythmias (atrial flutter or fibrillation) and the risk of pulmonary embolism, both being main causes of clinical and hemodynamic deterioration, a total cavopulmonary connection has been suggested with the aim of achieving a partial (anastomosis of the superior vena cava and pulmonary artery to the intra-atrial tunnel of the inferior vena cava) or complete (extracardiac conduit) bypass of the right atrium. A total cavopulmonary connection with an external conduit and without scarring of the parietal atrial wall significantly decreases the arrhythmic substrates of tachyarrhythmias[60]. The suitability for a total cavopulmonary connection is decided on the basis of Choussat's criteria. The introduction of the atrial septal fenestration technique with the aim of avoiding venous hypertension (> 14 mmHg) has resulted in an improved early postoperative hemodynamic status, but has not modified the long-term survival[61,62].

The conversion from an atriopulmonary connection to a total cavopulmonary connection together with the perioperative ablation of the reentry circuits seems to be effective in patients who do not respond to antiarrhythmic drugs[63]. Moreover, this technique may also be indicated as an alternative to cardiac transplant in patients who present with pleuropericardial or ascitic effusions and protein-losing enteropathy.

Correction of atrioventricular septal defects. In the last 30 years, the perioperative mortality in case of surgical correction of atrioventricular septal defects has decreased from 20-25% to 3-4%[64].

The mid- and long-term results were not so satisfactory because the patients have maintained a significant morbidity with a moderately high incidence of reoperation, mainly indicated because of the delayed onset of disorders of the atrioventricular conduction system, because of a residual intracardiac shunt and owing to incompetence of the left atrioventricular valve.

Comparison of the survival curves of different groups of patients operated upon for complete atrioventricular canal at different times does not reveal any significant modifications: the survival was 85% after 12-15 years of follow-up in the series of patients published by Studer et al.[65] and Daliento et al.[66] in 1982 and

1987 respectively; 81 and 90% survivals were reported after 8-10 years of postsurgical follow-up in the series of patients published by Ashraf et al.[67] and Tweddell et al.[68] in 1993 and 1996 respectively. Residual pulmonary hypertension, arrhythmias and heart failure distinguished the clinical history of patients who died during follow-up. An earlier surgical repair should help to limit delayed complications such as pulmonary hypertension but incompetence of the left atrioventricular valve remains the Achilles' heel affecting the postsurgical history and work capacity.

Women with congenital heart disease. More than 40% of GUCH patients are women. In females adolescence involves specific aspects related to menarche, procreation and sexuality[69]. As may be inferred from the data of a series of adults < 20 years old collected at the GUCH unit of Brompton Hospital in London in which the gender distribution was the same as in earlier life[70], the patient's sex does not seem to modify survival until adolescence and young adulthood. Nevertheless, even if among males the prevalence of aortic stenosis with or without coarctation or tetralogy of Fallot is not unexpected, it is surprising to find a higher percentage of young females presenting with Eisenmenger's reaction. All the same, in a series of patients older than 40 years, more males with Eisenmenger's reaction or tricuspid atresia or Fontan's operation were found. If we consider the main causes of death in adult patients with Eisenmenger's reaction, the relationships between a poor prognosis and pregnancy, extracardiac surgery prevalently due to gynecologic pathology and a higher incidence of cerebral abscesses probably related to paradoxical embolism, decompensated erythrocytosis and loss of iron with menstruation, the use of hormones for contraception or other gynecological problems are clear. Indeed, pregnancy is absolutely contraindicated in patients presenting with Eisenmenger's reaction. In our collaborative study[10], we noted 42 pregnancies in 24 women: only 10 in 8 patients continued beyond 26 weeks. In the latter cases, all pregnancies were delivered prior to 37 weeks of gestation and 3 women died within the first 10 days of delivery. Autopsy revealed widespread fibrinoid changes in the pulmonary arterioles with no evidence of pulmonary thrombi in the lungs. Clinical deterioration was found in all survivors. Moreover, pregnancies in patients with pulmonary atresia and an interventricular septal defect, univentricular and cyanotic heart disease, with a history including the Fontan or Mustard or Senning operations, severe obstruction of the left outflow tract, reduced systemic ventricular function secondary to a residual systolic overload or valvular incompetence or a degenerated homograft, mechanical prosthetic valve and Marfan's syndrome have to be considered high-risk gestations[71,72]. Aortic bicuspid valve, moderate stenosis of the aortic valve, stenosis of the pulmonary valve, atrial septal defects, restrictive ventricular septal defects, persistent ductus arteriosus, mitral valve prolapse, and operated tetralogy of Fallot are congenital heart defects for which pregnancy is to be considered as a low-risk event. Prepregnancy counseling and clinical evaluation are mandatory and should include physical examination, an accurate assessment of the hemodynamic status and functional capacity, arrhythmic risk stratification, evaluation of the maternal risk of morbidity and mortality, the impact of pregnancy on long-term survival and the risk that the fetus inherits congenital heart disease. The incidence of congenital heart disease in offspring ranges from 2 to 50% and it is higher when the mother, rather than the father, has cardiac disease[73]. The risk is the highest for single gene disorders and/or chromosomal abnormalities such as Marfan's syndrome, Noonan's syndrome and Holt-Horam's syndrome. Genetic counseling should include fetal echocardiography that can be performed at 16-18 weeks of gestation. The introduction of fetal echocardiography has had significant consequences in terms of the pregnancy outcome. In London and Paris more than 60% of pregnancies were interrupted after a fetal diagnosis of congenital heart defect[69]. On the contrary, in Italy there are no more than 20% of voluntary abortions. Socio-economic conditions, ethical and religious motivations and the modality of information determine the final decision women take. In England, for example, the mothers of babies with a hypoplastic left ventricle are exclusively adolescents or very young unmarried women of minority ethnic groups who received inadequate medical information[70]. The patient's decision was different when information about the natural and postsurgical history of cardiac defects was given by a cardiologist or cardiac surgeon. Cyanotic patients without pulmonary hypertension have a low risk of dying during pregnancy (only one death in the series of Presbitero et al.[74]; the patient, to whom no antibiotic prophylaxis was prescribed during delivery, died of endocarditis 1 month after delivery) but a very high risk of spontaneous pregnancy termination. The probability of success is < 10% when the aortic oxygen saturation is less than 80%. Moreover, contraception in patients at high risk creates problems. In our patients with Eisenmenger's reaction, of 9 who were on low estrogen pills, clinical deterioration including a worsening cyanosis has been observed: 2 of them had one episode of cerebral embolism resulting in one death. Two had undesired pregnancies; the contraceptive method used was the mini-pill in one case and the diaphragm in the other. One woman with an intrauterine device developed endocarditis. Twenty-two patients had elective sterilization and 2 died during surgery. The request for a surrogate pregnancy with another women using her own eggs had to be rejected because of the danger posed by the level of hormones required for oocyte harvesting and by the administration of general anesthesia.

Factors determining the quality of life

The conditions determining the quality of life and well-being are both objective, such as education, employment, family background and informed medical care, as well as subjective, such as self-perception and self-evaluation, and symbolic – those related to one's cultural background and to lifestyle of the social group to which he belongs. Often the normality acquired following surgery clashes with reality[75].

Each member of the GUCH medical community has a personal postsurgical history, depending on the anatomy of the native malformation, the hemodynamic overload and the surgical technique. It is a medical history which requires expert cardiological and surgical care, but it is also a biological history including puberty, sexuality, contraception, pregnancy and the risk of affected offspring. It is a social history including employment problems, the definition of the medications affecting the patient's performance or basic disability, legal problems involving life and medical insurance and psychological problems. These patients have many needs and many questions.

Not giving a correct answer to each specific question reduces the entity of surgical success.

Does the patient's care rightly belong to the pediatric cardiologist or to the cardiologist? A pediatric cardiologist, who knows the anomaly and the family, often wants to continue the care, but the latter is rarely satisfactory because he does not have access to adult beds and he has no knowledge of adult cardiac disease or about the medical management of adults. The majority of adult cardiologists are not culturally equipped to satisfy the increasing requests for care which are emerging from this new medical community. Moreover, the Division of Cardiology does not have space specifically reserved for these patients and GUCH units are few in number. The result is that many adult congenital heart disease patients receive suboptimal medical care and that for many the outcome may be unnecessary deterioration and even premature death[76].

The aim of both the pediatric and Adult Scientific Societies of Cardiology must be to organize the training of cardiologists for adult patients with congenital heart disease together.

Few but special care units for these patients have to be provided in the Division of Adult Cardiology where different experts (hemodynamists, electrophysiologists, pediatric cardiologists and cardiac surgeons, psychologists, anesthesiologists) need to collaborate.

The quality of life of these patients depends on the interaction between their clinical conditions, psychological state, environment and social and health services. Our contribution should be to help achieve a personal life project for these patients, stressing their abilities rather than their disabilities and paying more attention to how they perceive the normality of their lives.

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