Editorial Comment

Congenital Heart Disease in Adult Patients

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The number of adult patients with congenital heart disease is growing steadily. According to Monro,[1] it could reach 25% by the end of this decade, which means that in 2010 the patients undergoing surgery for congenital heart disease who are over 16 years old will outnumber those below that age. The incidence of congenital heart disease is of the order of 8-10 cases per 1000 births.[2] The natural history of these patients, regardless of the type of congenital heart disease and the method of treatment, has been analysed in previous studies. It has been reported that their mortality is fourteen, six and three times higher than that of the general population, for ages up to 14 years, 15-20 years and over 21 years, respectively. If the diseases are ordered according to their severity, the mortality is three times higher than in the general population for congenital heart diseases of mild degree, 13 times for moderate and 77 times higher in the most severe diseases.[3]

Nowadays, these data have changed, mainly as regards our diagnostic capabilities and our ability to discover congenital heart disease early on. Thus, in almost all cases a patient will undergo corrective or palliative surgery, according to the severity of the disease, as a neonate, infant or child. Corrective surgery is employed in the case of a pre-existing pathological condition (where there had never been a normal condition), which we attempt to make normal. With palliative surgery we try to improve the patient's clinical condition, while the underlying pathology remains. There is also surgical repair, when an attempt is made to restore a previously existing normal state. Obviously, the term "repair" cannot be used in the case of congenital heart disease: only the first two terms apply.[4]

If we exclude the successful closure of a secondary atrial septal defect, a ventricular septal defect, or a patent ductus arteriosus in early life, all the remaining congenital heart diseases will need care even in adulthood, since there may be expected to be significant residual lesions, relapses, other consequent disorders, or degenerative changes in prostheses, all of which will need to be dealt with at the appropriate time. The total population of patients with congenital heart disease who reach adulthood make up a new medical community, known internationally as "grown-up congenital heart patients" (GUCH). In this category we also have to include a number of adult patients who slipped through the net in earlier eras, survived because of the low or moderate severity of their congenital heart disease, and present for primary surgical treatment and care after they become adults. It is interesting to note that 30% of admissions of adult patients with congenital heart disease are over the age of 40 years and 5% are aged over 60.[5]

All the above, along with the increasing health care needs of specific categories

of patient, necessitate a change in our approach to the management of such cases. Their hospital treatment in most parts of the world takes place in cardiology departments dealing with adults. We believe that it is necessary to set up special, vertically organised congenital heart disease groups, staffed by paediatric cardiologists, cardiologists specialising in congenital heart disease, and cardiac surgeons specialising in the treatment of congenital heart diseases in patients of all ages. In view of this, we welcome the existence of such a group in Greece, the Department of Paediatric and Congenital Heart Surgery of the Onassis Cardiac Surgery Centre, whose experience with adults is described retrospectively elsewhere in this issue.[6]

The first thing to say about the study in question is that it is one among few that exist in the international literature. Overall, it is an extremely interesting study, since this is the first time in Greece that the results of corrective surgery for congenital heart disease in patients who managed to survive until adulthood, with or without previous surgical treatment of their condition, have been presented. Looking at the number of patients included in the study, we see that the activity of this centre in the treatment of adult patients with congenital heart disease (289 patients in seven years) exceeds that of a similar regional centre in Great Britain, such as the Birmingham Children's Hospital, which treated 474 patients in 13 years,[7] while it is not far behind the activity of an internationally recognised centre, such as the Congenital Heart Unit of the Royal Brompton Hospital and National Heart and Lung Institute in London, which reported treating 295 patients between 1990 and 1994.[8] The most important difference between the two latter centres is in the percentage of reoperations, which was 34.2% in Birmingham and 58% in London. The small number of reoperations in the Greek data (13.15%) is, I believe, largely due to the fact that Greek children with congenital heart disease used to be treated mainly in centres abroad, so that most of the reoperations that became necessary were also carried out in the same centres, for psychological reasons, or because they knew the history, and so on. A second reason is that in order for a centre of this type to create a patient population needing reoperation, it must be in existence for 20 years or more. The group presenting the study in question has existed for just seven years, so the small number of reoperations is to be expected. Indeed, in a personal conversation with one of the authors I learned that the majority of patients who presented for reoperation had been previously treated in another centre.

Of all the patients with congenital heart disease who undergo corrective surgery for the first time in adulthood, the lion's share in the international literature involves, in descending order of frequency, closing an atrial septal defect, aortic valve replacement, and coarctation of the aorta. This was verified in the Greek study, where the respective percentages were 54%, 19% and 6.4%. However, the number of atrial septal defects was impressively large (153 patients in seven years) if it is compared with the large, multicentre study, Pediatric Cardiac Care Consortium, which took place in the USA during 1984-1994 and included around 32000 patients of all ages who underwent surgery for congenital heart disease.[9] In that series the adults with an atrial septal defect numbered 203. This means that in Greece, until at least two decades ago, the diagnosis of atrial septal defect was deficient. I believe that in the future the number of adult patients with atrial septal defect will fall dramatically, as a result of the more thorough investigation of congenital heart diseases and their treatment during childhood, as well as the increasing use of invasive methods that close the defect with various types of "umbrella". This view is supported by other studies that have recorded a reduction in such cases by 60% during the last decade.[7] In such a large series of atrial septal defects in adults it would have been interesting to be given information about the postoperative arrhythmiological findings in those patients in relation to the age at which they were treated and their preoperative condition. It is well known that around half the patients with atrial septal defect and preoperative arrhythmia (atrial fibrillation or flutter) continue to have an atrial arrhythmia postoperatively, especially those over 40 years of age.[10]

The changes in the practical management of congenital heart disease in adults during recent years have mainly been expressed through an increase in reoperations, a reduction in cases with a "simple" diagnosis and an increase in the cases of replacement of pulmonary conduits in surgically treated tetralogy of Fallot.[7] Concerning the reoperations, we can divide them into those that are the first corrective procedure following an original palliative operation (mainly in aortic valve disease, correction of tetralogy of Fallot and congenital heart diseases that require a Fontan procedure), reoperations that involve

a new repair of an original corrective procedure (in the aortic valve, for right-sided dysfunctional conduits, restenosis in coarctation of the aorta, etc.), and reoperations that are further palliative procedures to supplement the original surgery.[8] In the Greek study published here six of the total of 36 reoperations refer to cases of tetralogy of Fallot, but the others are not specified.

In a multicentre study by the European Congenital Heart Surgeons' Association, which is the largest study published to date with 1247 adult patients who underwent surgery for congenital heart disease,[11] the 30-day mortality was 2.4%, with a range from 0% to 15.3% in the various centres. The 4-year survival was 94% overall and was higher in primary corrective procedures (95%) than in reoperations (92%) or in palliative procedures (88%). The results presented here from the Greek study are indeed exceptional, in that only one early death was recorded (which could perhaps have been usefully analysed in the study: diagnosis, procedure, outcome, postoperative day). As regards long term survival the results are also striking, with only two postoperative deaths. The 26 patients in NYHA functional class II and the single patient in class III obviously do not alter the authors' view that "All the remaining patients are in excellent clinical condition." Factors affecting mortality are cyanosis, the number of prior procedures and a patient's great age. A mortality rate of 18% has been described in cases of central cyanosis and one of 13% in patients who undergo more than two previous operations.[8,11]

Sudden cardiac death following surgery in adult patients with congenital heart disease is many times more common than in the general population. At a congress of the Canadian Cardiovascular Society in 1994 it was determined to occur in 5.3 out of every 1000 patients followed up annually. More specifically, of a total of 92 confirmed deaths 23 were sudden (24%), with nine of those patients having Eisenmenger syndrome, 12 a history of a corrective procedure, six palliative procedures and five never having undergone surgery. There was a previous history of ventricular arrhythmia in only three patients.[12]

The quality of life of adult patients with congenital heart disease has been the subject of a number of studies. The general perception is that all patients have a poorer level of physical activity than the general population. Cyanotic patients have a significantly poorer quality of life compared to patients of similar age and sex who have non-cyanotic disease. Patients who undergo a palliative procedure have the same quality of life as patients whose underlying disease does not require surgery. The surprising thing is that patients who have had a completely successful correction of an atrial septal defect show a significantly poorer quality of life, according to all parameters apart from pain, than does the general population.[13,14]

In closing, I would like to make three main observations. The first is that the number of adult patients with congenital heart disease will continue to grow. The second is that in this special group of patients there will be a need for more and more increasingly complex reoperations. The third is that there is a need for the creation of treatment centres for adult patients with congenital heart disease, whose specialisation can lead to higher standards of performance. The retrospective study by Sarris' group in this issue makes a rich contribution to the rather poor literature and its findings emphatically confirm the value of special centres for congenital heart disease.

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