State of the Art

Cardiac Surgery in Adults with Congenital Heart Disease: An Emerging Challenge

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Progress in pediatric cardiac surgery over the last 20 to 30 years has allowed a large number of neonates, infants and children with congenital heart diseases to survive until adulthood. As a result, while in former times the term "congenital heart disease" applied mainly to childhood (since adults with congenital heart disease, then encountered only rarely, usually had fewer complications and a relatively benign natural history), today it is estimated that the steadily growing number of adults with congenital heart diseases has probably overtaken the number of child patients. The adult population with congenital heart disease (grownup congenital heart disease) has important peculiarities and requirements. The spectrum of anomalies and the complexity of the postoperative anatomy and pathophysiology may extend beyond the training and experience, not only of most cardiologists who deal with adults, but also of cardiac surgeons who are involved only with general cardiac surgery in adults. On the other hand, pediatric cardiologists who wish to continue to follow their patients as they grow into maturity often have difficulties when they are asked to treat acquired heart conditions and other adult diseases that afflict those patients. Similar problems are faced by pediatric cardiac surgeons who have given up surgery in adults—for example, as a result of working exclusively in pediatric centers. Consequently, there is an increased need for centers that bring together the skills of cardiac surgeons and cardiologists or pediatric cardiologists who have knowledge and experience of congenital heart disease in adults. Recognizing the importance of this problem, at its 32nd Meeting in Bethesda (2-3 October, 2000) the American College of Cardiology endorsed an investigation into the subject: "Care of the Adult with Congenital Heart Disease." The associated report was published in the College's journal in the spring of 2001.[1]

Epidemiology

It is estimated that the number of adults with congenital heart disease in the USA has now reached 800,000, not including patients with isolated bicuspid aortic valve.[1] Of those, around half have quite complex diseases requiring follow up and treatment by specialized cardiologists and cardiac surgeons. Since we have no studies of the Greek population from which to draw relevant statistical data, we can only make a proportional approximation that there are around 15,000 adults in Greece who suffer from significant congenital heart disease and need follow up and treatment by specialists.

According to the estimates of the American College of Cardiology, the number of adults with congenital heart disease must now exceed the number of children. For example, since the great majority of patients

Table 1. Diagnoses of adult patients with complex congenital heart diseases. These patients need regular follow up in specialized centers. (Modified from the 32nd Bethesda Conference.[1])

1. Truncus arteriosus / semitruncus
2. Mitral valve atresia
3. Pulmonary valve atresia (all forms)
4. Tricuspid valve atresia
5. Double-outlet ventricle
6. Fontan procedure
7. Cyanotic congenital heart disease (all forms)
8. Single ventricle (double inlet or double outlet, common or primitive ventricle)
9. Transposition of the great vessels
10. Conduits (valve-bearing or not)
11. Pulmonary vascular obstructive disease
12. Other atrioventricular or ventriculoarterial connection anomalies (e.g. criss-cross heart, isomerism, heterotaxy, ventricular inversion)

with the serious defect of transposition of the great vessels now survive, thanks to the corrective Mustard, Senning and arterial switch procedures, it was estimated in the year 2000 that most of the children with transposition of the great vessels who were being followed in the Hospital for Sick Children in Toronto will be adults after 2008.[2]

Categorization of patients

Adults with congenital heart disease can be divided into three categories: those with complex heart diseases (Table 1), those with diseases of moderate severity (Table 2), and those with relatively simple defects (Table 3). It should be noted that the numbers of adults with congenital heart disease quoted above underestimate reality. This is partly because they are based on the rates of appearance of congenital heart diseases in infancy and childhood, whereas at least 10% of adults with congenital heart disease sent for treatment in most of the specialized centers suffer from conditions that have been diagnosed for the first time after they reached adulthood, such as atrial septal defect, Ebstein's disease or corrected transposition of the great vessels.

Adults with congenital heart disease who fall into the first two categories (moderate severity or complex diseases) must be followed at regular intervals and

Table 2. Diagnoses of adult patients with congenital heart diseases of moderate complexity. These patients need regular follow up in specialized centers. (Modified from the 32nd Bethesda Conference.[1])

1. Aneurysm or fistula of the Valsalva sinus
2. Patent ductus arteriosus
3. Anomalous pulmonary venous return, partial or full
4. Aortic-left ventricular tunnel
5. Aortic isthmic coarctation
6. Atrioventricular canal defects, partial or full
7. Atrial septal defect, sinus venosus type
8. Ventricular septal defect with:
   - Missing valve
   - Aortic regurgitation
   - Aortic isthmic coarctation
   - Mitral valve disease
   - Stenosis of the right ventricular outflow tract
   - Straddling mitral or tricuspid valve
   - Aortic subvalvular stenosis
9. Ebstein's anomaly
10. Significant pulmonary regurgitation
11. Significant pulmonary valve stenosis
12. Stenosis of the right ventricular outflow tract
13. Tetralogy of Fallot
14. Aortic subvalvular or supravalvular stenosis (apart from hypertrophic obstructive cardiomyopathy)

Table 3. Diagnoses of adult patients with relatively simple congenital heart diseases. These patients may be followed up in non-specialized centers, provided an initial full examination has been carried out in a specialized center. (Modified from the 32nd Bethesda Conference.[1])

Uncorrected defects

1. Lone aortic valve disease
2. Lone mitral valve disease (apart from "parachute valve", "cleft leaflet")
3. Lone patent foramen ovalis without complications
4. Lone small (hemodynamically insignificant) ventricular septal defect
5. Mild pulmonary valve stenosis

Following surgical correction

1. Patent ductus arteriosus
2. Atrial septal defect (without residual defect)
3. Ventricular septal defect (without residual defect)

treated by physicians (cardiologists, cardiac surgeons) with special training and experience in congenital heart disease, preferably in specialized centers. Defects that can also be monitored by physicians without special training in congenital heart diseases, outside specialized centers, are summarized in table 3 and include simple valvular diseases as well as corrected small atrial or ventricular septal defects. However, in any case it is recommended that the follow up of all patients with congenital heart disease should be coordinated by a specialized center. It has been estimated that one center for adults with congenital heart disease is needed per 5-10 million population. The essential requirements for such a center are summarized in table 4.

The role of cardiac surgery

In the international literature there are not many large series of adult patients undergoing cardiac surgery for congenital heart disease.[4-6] In a series of 112 procedures in adults with congenital heart disease at Baylor College of Medicine – Texas Children's Hospital[5] the surgical mortality was 6% and the late mortality 3%. The recent multi-center study organized by the European Congenital Heart Surgeons' Association (in which our Department was an active participant and contributor of data) analyzed the results of the surgical treatment of adults with congenital heart disease in relation to 1342 patients in 19 participating European centers.[6] The overall mortality was 2.4%. Almost 80% of the procedures were corrective (40% atrial or ventricular septal defects); almost 20% involved various right-heart defects, such as reoperation for tetralogy of Fallot; 18% were left-heart defects, 4.7% various palliative procedures (including procedures for a single ventricle) and 17.2% were repeat procedures.

Surgery for adults with congenital heart disease in Greece

Since 1997, when our Department began operation, we have operated on more than 1500 children with

Table 4. Requirements for the operation of a grownup congenital heart disease center. (Modified from the 32nd Bethesda Conference.[1])

1. Cardiac surgery for congenital heart disease in adults and children
2. Specialized (pediatric) anesthesiology
3. Cardiac surgery intensive care unit specialized in congenital heart disease
4. Cardiology (adults and children)
   a. Diagnostic and invasive hemodynamic laboratory
   b. Echocardiography
   c. Electrophysiology
5. Imaging department (CT, MRI)
6. Specialized nursing personnel
7. Possibility of counseling in high-risk pregnancy

diagnoses covering the entire spectrum of congenital heart disease, with an overall mortality of <2%. Even though most of those children have still not reached adulthood, our Department has also been a focus of attraction for adult patients with congenital heart disease. These patients have either been operated on in other centers (mainly abroad) or were referred with a new diagnosis or new indications for surgical treatment of their congenital heart disease. This very positive experience, with surgical mortality 0.34%, is reported elsewhere in this issue[7] and confirms the need for these patients to be treated in special centers in order that the best possible outcome may be achieved. It should be noted that, apart from their referral to special centers, the increase in cases of grownup congenital heart disease in Greece has created a need for the creation and maintenance of a detailed database for their long term follow up. Our Department maintains such a database in collaboration with the European Congenital Heart Surgeons' Association, aimed at quality control and a continual improvement in results.

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