Nowadays, it is estimated that 85% of the infants born with congenital heart disease (CHD) will survive to adulthood, thanks mainly to surgical or therapeutic procedures performed during infancy or childhood. The clinical profile and disease pattern of adults with CHD is changing. The prevalence of certain adult CHDs, such as tetralogy of Fallot, transposition of the great arteries or univentricular heart, is rising, but these conditions have practically become new diseases as a result of therapy. Most surviving patients present residua, sequelae, or complications, which can progress during adult life. These disorders can present electrophysiological disturbances, valvular disease, persistent shunts, myocardial dysfunction, pulmonary or systemic vascular disease, problems caused by prosthetic materials, infectious complications, thromboembolic events, or extravascular disorders involving multiple organs or systems. In tetralogy of Fallot, the most striking problems that affect long-term prognosis are pulmonary valve regurgitation, right ventricle dysfunction, and atrial or ventricular arrhythmias. The main problems appearing after physiological atrial repair of transposition of the great arteries are related to right ventricular function, since it is structurally unprepared for systemic circulation, and atrial arrhythmias. Surgical repair of univentricular heart using Fontan techniques should be considered a palliative procedure that does not modify the underlying structural disorder and exposes the postoperative patient to severe complications and problems. The increase in the number of patients with CHD who will reach adulthood in the coming decades makes it necessary to carefully consider the new healthcare demands that are being generated, who should be responsible for them, and how and where solutions can be found.

Key words: Congenital heart disease. Cardiac surgery. Tetralogy of Fallot. Transposition of the great arteries. Univentricular heart.

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Abbreviations

CHD: congenital heart disease.
ASD: atrial septal defect.
VSD: ventricular septal defect.
TF: tetralogy of Fallot.
TGV: transposition of great vessels.
ACHDU: adult congenital heart disease unit.

Sequela, and complications of CHDs treated surgically in childhood. Undoubtedly, the impressive progress of cardiac surgery and advances in pediatric cardiology achieved in recent decades have been responsible for this evolution.1

Historical Perspective

From 1938, when Robert Gross, a pediatric surgeon of Boston, performed the first ligation of a ductus arteriosus in a 7 1/2 year old girl,2 to 1981, when Norwood3 devised a systematic approach to the surgical correction of aortic atresia, surgical techniques have been designed to correct, or at least palliate, any type of CHD not associated with unviable chromosomal or systemic anomalies (Table 1). The first achievements in this endeavor were made by Clarence Crafoord, who conceived the idea for the surgical correction of aortic coarctation,4 Alfred Blalock, who performed the first connection of the left subclavian artery to the ipsilateral pulmonary artery in cyanotic patients with pulmonary stenosis or atresia,5 and Richard Brock, who carried out the first pulmonary valvulotomy using a closed technique.6 At the beginning of the 1950s, Gibbon perfected a machine for extracorporeal circulation,7 which made it possible to operate directly on the heart.

The main achievements of the 1950s were the repair of ventricular septal defect (VSD) and tetralogy of Fallot8 (TF); of the 1960s, atrial physiological correction of transposition of the great vessels9 (TGV) and the interposition of conduits between the right ventricle and pulmonary artery;10 and in the 1970s, the techniques of Fontan11 and Jatene12 for the reconstruction of single-ventricle heart and TGV, respectively. The progressive improvement in cardiopulmonary bypass techniques, and the use of methods for cerebral13 and myocardial protection,14 have made the surgical repair of most CHDs possible at a progressively earlier age, obtaining excellent short and long-term results. The first successful heart transplantation in newborns was carried out at the Loma Linda university medical center in 1985.15 Since then, many centers in the world have offered this therapeutic alternative for infants with CHD unsuitable for surgical reconstruction or in which surgery has failed.

The initial contribution of pediatric cardiology was the morphological analysis of all the cardiac malformations. A select group of cardiologists and anatomists — notably Van Praagh16 in the U.S., Anderson17 in Great Britain, and Quero-Jiménez18 in Spain — dedicated considerable effort to the sequential analysis, nomenclature, classification, and diagnostic and therapeutic approach to CHD. The second advance was the use of cardiac catheterization and angiography techniques in the pathophysiological and anatomic assessment of CHD.19,20 In the 1980s, the hemodynamic and morphological assessment of CHD with non-invasive techniques began, and included techniques such as Doppler21 echocardiography,21 radionuclide angiography,22 and magnetic resonance imaging,23 thus making possible a broad array of therapeutic interventions in the catheterization laboratory. The first intracardiac therapeutic procedure carried out with a catheter was atrial balloon septostomy, which was made by Rashkind in 1966,24 and eventually led to

Table 1. Development of cardiac surgery in congenital heart diseases

<table>
<thead>
<tr>
<th>Year</th>
<th>Author</th>
<th>Surgical technique</th>
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<tbody>
<tr>
<td>1938</td>
<td>Gross</td>
<td>Ductus arteriosus ligation</td>
</tr>
<tr>
<td>1944</td>
<td>Crafoort</td>
<td>Aortic coarctation</td>
</tr>
<tr>
<td>1945</td>
<td>Blalock</td>
<td>Subclavian-pulmonary fistula</td>
</tr>
<tr>
<td>1948</td>
<td>Brock</td>
<td>Pulmonary valvulotomy</td>
</tr>
<tr>
<td>1953</td>
<td>Lillehei</td>
<td>VSD closure</td>
</tr>
<tr>
<td>1955</td>
<td>Kirklin</td>
<td>Fallot correction</td>
</tr>
<tr>
<td>1964</td>
<td>Mustard</td>
<td>Physiological TGV correction</td>
</tr>
<tr>
<td>1967</td>
<td>Rastelli</td>
<td>Prosthetic conduits</td>
</tr>
<tr>
<td>1971</td>
<td>Fontan</td>
<td>Atrialpulmonary connection</td>
</tr>
<tr>
<td>1975</td>
<td>Jatene</td>
<td>Anatomical TGV correction</td>
</tr>
<tr>
<td>1981</td>
<td>Norwood</td>
<td>Hypoplastic left ventricle</td>
</tr>
<tr>
<td>1985</td>
<td>Fricker</td>
<td>Heart transplantation in children</td>
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</table>

Table 2. Therapeutic procedures in congenital heart disease

<table>
<thead>
<tr>
<th>Procedure</th>
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<tr>
<td>Atrial balloon septostomy</td>
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<tr>
<td>Atrial blade septectomy</td>
</tr>
<tr>
<td>Embolization of arteriovenous fistulas</td>
</tr>
<tr>
<td>Extraction of intravascular foreign bodies</td>
</tr>
<tr>
<td>Pulmonary or aortic valvuloplasty</td>
</tr>
<tr>
<td>Angioplasty and stenting of a native coarctation or re-coarctation</td>
</tr>
<tr>
<td>Angioplasty and stenting of pulmonary arteries</td>
</tr>
<tr>
<td>Dilatation of caval or pulmonary veins</td>
</tr>
<tr>
<td>Angioplasty and stenting of intra-atrial baffles</td>
</tr>
<tr>
<td>Occluder device for ductus arteriosus, ASD, and VSD</td>
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<tr>
<td>Percutaneous pulmonary prosthesis</td>
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</table>
the development of the present therapeutic arsenal available to pediatric cardiologists. The latest contribution of pediatric cardiology is prenatal diagnosis with fetal echocardiography that, without doubt, is an extraordinary advance in the knowledge of morphological and functional disturbances of the heart from an early gestational age.

**DEMOGRAPHIC IMPACT**

The major development experienced by pediatric cardiology and cardiovascular surgery in recent decades has made possible an impressive recovery program for children who once were condemned to die. The demographic impact of this advance has been reviewed in recent publications. In 1992, Moller and Anderson reviewed the present state of 1000 consecutive unselected children with CHD who were initially evaluated between 1952 and 1963. After a follow-up of 26 to 37 years, 285 patients had died, but 712 (71%) remained alive, most of them in an excellent or good clinical situation. In 1993, the findings of a prospective study on the long-term evolution of pulmonary stenosis, aortic stenosis, and VSD were published. After more than 25 years of follow-up, 97% of the patients with pulmonary stenosis, 87% of the patients with aortic stenosis, and 85% of the patients with VSD remained alive, most of them with a good or excellent functional assessment. The patients who underwent surgery for uncomplicated atrial septal defect (ASD) in the first two decades of the life had a long-term survival similar to that of the general population. The survival of patients with TF 30 years after surgery is 86%. Long-term follow-up studies of patients operated for complex heart diseases are less definitive, but in a recent study of 216 patients with tricuspid atresia, corrected with Fontan technique and followed-up for 25 years, the overall survival was 79%. Nowadays, it is estimated that 85% of the children born with CHD survive until adulthood and it is estimated that this percentage will increase even more over the next two decades, due to the continuous improvement in surgical and therapeutic techniques. In industrialized countries with a low birth rate, the number of adults with CHD now equals that of children with cardiac malformations. A conservative estimate of the number of patients with CHD who have survived to adulthood in the U.S. yields the figure of 787 800 cases for the year 2000, of which 117 000 had severe CHD and 302 000 CHD of moderate severity. By extrapolating these findings to the Spanish population, it is estimated that there are more than 100 000 adults with CHD in the country.

In order to attend this increasing demand, in recent years clinical units specialized in adult CHD (ACHDU) have been created in major health centers in the world. Spain, particularly the La Paz University Hospital of Madrid, has participated in this development. This center harbors a very active nucleus of pediatric cardiology and cardiac surgery, which has been functioning for more than 35 years. In this time, thousands of children with CHD have been studied and treated in the children’s hospital and many of them are now adult patients under clinical follow-up in the general hospital. According to mutual agreement between pediatric and adult cardiologists, in 1987 it was decided to unify the clinical follow-up.

![Fig. 1. Distribution by age groups of 2169 patients followed up in the Adult Congenital Heart Disease Unit the La Paz Hospital between 1990 and 2000.](image-url)
of these patients in an ACHDU. Between 1990 and 2000, 2169 adults with CHD were cared for in this unit. As can be seen in Figure 1, the number of patients over the age of 40 years who are cared for has remained stable, but there has been an avalanche of new cases in patients between the ages of 15 and 40 years, the majority coming from the children’s hospital. The analysis of this population is the main objective of this article.

CLINICAL FORMS OF SURVIVAL

The population of adult patients with CHD currently includes two large groups: a) CHD with natural survival and b) CHD with some type of therapeutic intervention (surgery or a percutaneous procedure) carried out in childhood. Cases of CHD with a natural survival until adulthood can be classified in three subgroups:

1. CHDs that require a primary therapeutic intervention in adulthood.
2. CHDs that do not require therapeutic intervention but do need monitoring and continuous clinical follow-up.
3. CHDs that are currently inoperable, can only be treated by transplantation, or in patients in which the surgical risk is greater than the risk of the natural evolution (Table 3).

In Figure 2A are shown the main CHDs of the series of the La Paz Hospital that required a primary intervention during adulthood. Without doubt, ASD continues to be the most frequent disorder in this group, followed by valvular or subvalvular aortic stenosis, and uncomplicated aortic coarctation. Other CHDs, like pulmonary stenosis, ductus arteriosus, and VSD, are now treated surgically in adults with much less frequency. In fact, except for aortic stenosis, which can continue to evolve throughout life, most patients in this group must undergo surgery before reaching adulthood, which is why the number of primary interventions of CHD in older patients is decreasing.

Figure 2b shows acyanotic CHDs that have not required any intervention in our series. This group is numerous and important, and is prominently represented by patients with bicuspid aortic valve, restrictive VSD, and mild pulmonary stenosis. They are asymptomatic patients with mild heart diseases that do not require treatment but are exposed to serious complications, such as infectious endocarditis or progressive valve degeneration (stenosis or insufficiency).

In Figure 2c are shown the cyanotic CHDs in our series that have a natural survival until adulthood. In this group, Eisenmenger syndrome, single-ventricle heart, Ebstein anomaly, and pulmonary atresia with VSD are noteworthy. These patients are symptomatic

Table 3. Classification of adult congenital heart diseases

<table>
<thead>
<tr>
<th>Natural survival</th>
<th>Operation in adulthood</th>
<th>Follow-up without surgery</th>
<th>Inoperable except for organ transplantation</th>
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</thead>
<tbody>
<tr>
<td>Operation in childhood</td>
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and have serious heart diseases, but cannot undergo surgery with a reasonable risk. In some cases, cardiopulmonary transplantation, or isolated pulmonary transplantation with correction of the cardiac anomaly are the only possible options, but the risks of these therapeutic alternatives are almost always higher than those of the natural evolution. These patients can present serious complications: hemorrhage (profuse hemoptysis), infection (endocarditis, cerebral abscess), arrhythmia (atrial flutter or fibrillation), or thromboembolism. However, these patients generally remain relatively stable, with intolerance to effort, frequent symptoms of hyperviscosity, but few signs of cardiac insufficiency. In our experience, most complications occur during pregnancy, intercurrent disease, surgical interventions, impaired hydration, excessive exercise, or medical interventions. These patients need strict cardiological control to prevent high-risk situations and ensure that immediate treatment will be provided if complications arise: embolization of bleeding vessels, surgical drainage of cerebral abscesses, arrhythmia ablation, and others.

CHANGES IN THE SPECTRUM OF CONGENITAL HEART DISEASE IN ADULTS

Currently, the most numerous group of patients with CHD who reach adulthood is formed by patients who have undergone some type of therapeutic intervention during childhood. This fact is greatly changing the spectrum of CHDs that cardiologists see in adults, which underlines that cardiologists should be prepared to recognize this new disease. In the series of La Paz Hospital, 56% of adults under the age of 40 years pertained to this group. In Figure 3 are shown the main CHDs treated surgically in childhood in our series. Without doubt, the most frequent CHD in this group is TF. This disease had disappeared almost completely from general cardiology clinics and now has reappeared with a much greater prevalence, but in the form of a postoperative disease. We are also observing the appearance in our clinics of diseases that are practically unknown to us, like TGV. Complete transposition is an unviable CHD outside the first months of life and must be repaired by surgery. Nevertheless, most children with this malformation now reach adult life, although in a state modified by different surgical techniques. Patients with an anatomically or functionally unique heart constitute another large group of patients that are seen in our clinic. Most of them arrive after undergoing surgical palliation or partial correction, which enables survival but does not modify the basic structural disturbance and adds structural changes introduced by surgery.

In spite of the previous intervention, the patients in this group rarely reach adulthood with a fully normal anatomy, physiology, and cardiovascular function. Few therapeutic interventions are totally curative. The ligation of a ductus arteriosus, closure of an ostium secundum type ASD at an early age, or closure of a VSD by transatrial approach can be totally curative. In most cases, surgical or interventional procedures are only reparative and in other cases are just palliative in nature. This means that almost all patients with CHD who were operated during childhood present residual injuries, sequelae, or complications that can have a progressive character during adulthood.

RESIDUA, SEQUELAE, AND COMPLICATIONS

Table 4 offers a classification of residual lesions, sequelae, and complications of CHD in adults, modified from the Perlof and Warnes classification. These injuries can manifest as electrophysiological disturbances, valve disease, persistent shunting,
myocardial dysfunction, vascular lesions of the pulmonary or peripheral system, problems derived from prosthetic materials, infectious complications, thromboembolic phenomena, or extravascular disturbances that affect multiple organs and systems. In general, these problems can be caused by intrinsic malformations, the effects of therapeutic procedures, secondary hemodynamic changes, or unexpected complications. Residual lesions are anomalies that are deliberately left uncorrected during surgical repair. Examples of residual lesions are a restrictive VSD associated with a repaired aortic coarctation, or mitral insufficiency due to a cleft anterior leaflet that was not repaired during the surgical closure of an ostium primum type ASD. Neurological or systemic abnormalities that remain after the intervention are also residual lesions. Sequelae are new cardiovascular disturbances that occur as a necessary consequence of repair. Pulmonary failure secondary to the surgical correction of TF with a transannular patch, or persistent electrophysiological anomalies after right ventriculotomy are common examples. In contrast, complications are undesired cardiovascular or systemic disturbances that may be related to therapeutic procedures or appear spontaneously in the course of the natural evolution. Postoperative AV block, myocardial dysfunction due to insufficient protection of the cardiac muscle during ischemia, or the consequences of infectious endocarditis are complications that can have a major impact on survival and morbidity in adulthood. This review does not allow an exhaustive analysis of the residual lesions, sequelae, and complications of CHDs operated in childhood.44 However, in the following sections we will review the main residual disorders and the most frequent complications of patients with TF, TGV, and single-ventricle heart, three CHDs that have emerged in the context of adult heart disease as modified conditions that manifest as practically new diseases.

**TETRALOGY OF FALLOT**

TF is the most frequent cyanotic CHD in children.45 Fewer and fewer patients with this CHD reach adulthood by natural survival, but a growing number of adults have TF in postoperative follow-up. The estimated survival rate 30 years after surgery is 86%.44 Surgical correction consists of VSD closure and enlargement of the right ventricular outflow tract. For this reason, the surgeon often must use a transannular patch that can reach the pulmonary bifurcation and, sometimes, a prosthetic conduit between the right ventricle and pulmonary artery. It is not uncommon that the pulmonary branches have to be enlarged if there is congenital stenosis or stenosis induced by previous surgical fistulas.

**Obstruction of the outflow tract**

When they reach adulthood, most patients maintain an effective correction. In more than 60% of our series, the systolic pressure of the right ventricle is less than 50 mm Hg and there is no significant residual gradient, although 10% to 15% present moderate or severe obstruction of the outflow tract with a right ventricular systolic pressure that is more 70% greater than systemic pressure.44 The obstruction can be subvalvular, valvular, or supravalvular, although it is most frequently located in the branches of the pulmonary artery. This raises serious diagnostic problems in adults because the pulmonary branches are poorly evaluated by conventional or transesophageal Doppler echocardiography. Nevertheless, vascular magnetic resonance imaging is a high-resolution non-invasive technique for the anatomic assessment of the main pulmonary branches (Figure 4). The surgical treatment of stenosis of the pulmonary branches also

### Table 4. Classification of the residual lesions, sequelae, and complications of congenital heart disease

| Classification                |  
|-------------------------------|--------------------------------------------------|
| Electrophysiological disturbances | Permanent electrophysiological changes  |
|                               | Arrhythmias and conduction defects              |
| Cardiac valve disease         | Intrinsic valve malformation                    |
|                               | Sequelae of previous interventions             |
|                               | Hemodynamic effects on normal valves           |
| Persistent shunts             | Uncorrected residual lesions                   |
|                               | Sequelae of therapeutic procedures             |
| Myocardial dysfunction        | Structural disturbances                         |
|                               | Hypertrophy and remodeling                     |
|                               | Perioperative ischemia                          |
| Vascular disorders            | Congenital or acquired stenoses                |
|                               | Pulmonary or systemic hypertension             |
|                               | Aneurysm, dissection or rupture                 |
| Prosthetic materials          | Patches, valves, or conduits                   |
| Infectious complications      | Cardiac valves or mural endocardium            |
|                               | Arterial vessels or fistulas                    |
|                               | Extravascular structures (cerebral abscess)     |
| Thromboembolic phenomena      | Intravascular thrombosis                       |
|                               | Pulmonary or systemic thromboembolism          |
| Extravascular disturbances    | Mental and physical development                 |
|                               | Sense organs                                    |
|                               | Osteomuscular structure                         |
|                               | Central nervous system                          |
|                               | Teeth                                           |
|                               | Other organs and systems                        |

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has serious difficulties, but percutaneous procedures using stents are very effective\textsuperscript{46,47} (Figure 5). In patients who have undergone correction with a conduit or valve prosthesis, restenosis is the rule and many patients need to be reoperated several time in the course of their lives. Dilation with stent placement\textsuperscript{48} and later percutaneous implantation of a valve prosthesis is a new procedure that shows great promise in these patients.\textsuperscript{49}

### Persistent shunts

Residual shunts are not infrequent. Thirty percent of the patients in our series have a persistent shunt, in most cases small and of scant clinical importance.\textsuperscript{44} The most frequent location is the interventricular patch, although some patients have an extracardiac shunt related with surgical fistulas or unligated aortopulmonary collateral vessels. Some of our patients have a permeable foramen ovale or an ASD not closed in the correction. This has been cause of paradoxical embolism in some cases, which could have been avoided if more attention had been given to the interatrial septum during surgery. Most residual interatrial or aortopulmonary shunts can be treated with percutaneous procedures.

### Valvular and myocardial dysfunction

Some adult patients with operated TF develop progressive aortic insufficiency. Dilation of the aortic root, lack of support due to a septal defect, and valvular deformation resulting from retraction of the surgical patch are the main causes.\textsuperscript{50} Eight patients in our series were operated on as adults for severe aortic insufficiency. Nevertheless, the most frequent valve problem in patients who have undergone surgery for TF is pulmonary valve insufficiency. Approximately half of the patients in our series had grade III-IV pulmonary valve insufficiency due to the use of transannular patches to enlarge the ring, which leave the valve completely incompetent. Pulmonary insufficiency is practically a new disease, which is almost unknown as a natural disease. At present we are learning to recognize and assess it.\textsuperscript{51-53} We know that it is clinically well tolerated during decades, but it causes progressive expansion of the right ventricle. When the ventricular dilation is excessive, the increased postload limits cardiac output with exercise and the functional capacity decreases. With time, ventricular function deteriorates and the patient characteristically presents a hypotensive response to effort and a greater functional limitation.\textsuperscript{54,55}
ventricular dilation and dysfunction causes insufficiency of the tricuspid valve and increased pressure in the right atrium and caval veins. In the terminal stages, signs and symptoms of right heart failure appear.  

Arrhythmias and sudden death

Adults with operated TF experience frequent episodes of ventricular tachycardia or atrial tachyarrhythmia (flutter or fibrillation). These arrhythmias sometimes induce syncope or sudden death. A linear incidence of sudden death of 0.4% annually during the first 25 years after the intervention has been estimated, and the incidence grows exponentially from the first 25 years on. The arrhythmia substrate may be related to surgical scars, although the dilation of the right ventricle and atrium induced by the pulmonary and tricuspid insufficiency acts as a trigger factor. It has recently been confirmed that the patients with the greatest risk of atrial and ventricular arrhythmias, including sudden death, are those that have severe pulmonary and/or tricuspid insufficiency and develop a marked cardiomegaly, progressive widening of the QRS complex, and an increase in the dispersion of the QT interval. A cardiothoracic index >60%, QRS duration >180 ms and QT dispersion >60 ms are markers of risk that are easy to obtain at the bedside of the patient.

Reintervention in adulthood

Very recent findings suggest that surgical reintervention to correct pulmonary insufficiency with a valvular prosthesis and tricuspid insufficiency by annuloplasty decreases the incidence of atrial and ventricular arrhythmias, especially if surgical repair is accompanied by ablation of the reentry circuit. Unfortunately, we do not yet have trustworthy clinical markers to determine which asymptomatic or minimally symptomatic patients can benefit from early reintervention, but the general impression is that the intervention is usually too late. In most cases, the dilation and systolic dysfunction of the right ventricle do not remit after surgery. In the same way that we know the limits of ventricular size that allow functional normalization in the postoperative period of aortic and mitral insufficiency, we need to know what clinical and echocardiographic data are related to the normalization of right ventricular function after surgery.

Transposition of great vessels

TGV is the most frequent cyanotic heart disease at birth, but natural survival to an adult age is exceptional. This malformation represents 5%-8% of all CHD, but when left to their natural evolution, 30% of the patients die in the first week of life, 50% in the first month, 70% in the first 6 months of life, and 90% in the first year. Only one patient in our series has survived to adulthood without any type of intervention. Atrial septostomy with the Rashkind technique and atrial physiological correction with the Senning or Mustard techniques have radically changed the life expectancy of these patients. In the last decade, anatomical correction (arterial switch) is being used with increasing frequency, although its impact on long-term survival remains to be seen.

Dysfunction of the systemic right ventricle

Patients with TGV have a special anatomic situation in which the right ventricle is connected to the aorta and must bear systemic circulation. The use of the Mustard and Senning techniques for atrial physiological repair over a period of more than 20 years means that many patients have a right ventricle that is surgically connected to the aorta. In the postoperative follow-up, a large number of patients present systolic dysfunction of the right ventricle at rest or after physical exercise. As adults life, most of them are clinically well, but have a reduced capacity for effort. Some develop chronic heart failure and the proportion of patients with this complication increases with time. The cause of postoperative right ventricular dysfunction is multifactorial. The factors that may contribute are: a) inability of the right ventricle to adapt to its new function as the pump for systemic circulation; b) coronary circulation maintained by a right coronary artery that must supply all the blood flow to the systemic ventricle; c) myocardial damage produced by sustained severe hypoxia before surgery, and d) perioperative damage induced by insufficient myocardial protection.

Cardiac rhythm disorders

The high frequency of postoperative arrhythmias after atrial physiological correction is, along with myocardial dysfunction, one of the main reasons why this intervention is being replaced by arterial switch (Jatene) in many centers. The incidence of significant arrhythmias increases progressively with the time since the intervention. It has been estimated that the probability of maintaining a normal sinus rhythm is 72% at 1 year of the Mustard operation, 56% at 5 years, and 43% at 13 years. The most frequent rhythm disorders are symptomatic sinus bradycardia, slow nodal rhythms, and paroxysmal or sustained supraventricular tachycardia, principally atrial flutter. The cause lies in the lesion of the sinus
node during surgery and the atrial scars that interrupt the preferred internodal conduction pathways, which form the substrate for reentry circuits.71 The current indications for a permanent pacemaker in patients with TGV are complete AV block, sustained heart rates of less than 30 beats/min, documented Stokes-Adams episodes, severe ventricular systolic dysfunction with bradycardia, and the need to begin treatment with antiarrhythmic drugs that depress sinus function even more. In patients with recurrent atrial flutter, ablation of the arrhythmia may be indicated, although this procedure involves difficulties because the circuit it is located in the pulmonary venous canal in most cases, which makes a retrograde arterial approach mandatory.

Dehiscence and obstruction by baffles

These are the two most frequent mechanical complications that appear after atrial physiological correction with the Mustard or Senning technique. The obstruction can be early, due to an unsuitable reconstruction geometry, but it may also be late, due to retraction and calcification of the baffles and scars on the atrial wall. Dehiscence and baffle obstruction are usually located distal to the inflow tract of the superior vena cava.72 In small children, these defects can be easily diagnosed by color Doppler echocardiography,73,74 but in the postoperative adult patient, multiplane transesophageal echocardiography provides more diagnostic information (Figure 6). Obstruction of the inferior vena cava is, fortunately, less frequent, but it can induce Budd-Chiari syndrome, congestive liver disease, and protein-losing enteropathy.75 Baffle stenosis can be corrected by balloon dilation76,77 or an intravascular stent.78,79

Valvular dysfunction

Insufficiency of the tricuspid valve, which is located on the systemic ventricle, can appear after surgical atrial correction, but it is infrequent in the absence of severe right ventricular dysfunction. This complication is more prevalent in patients with a large VSD that has been closed with a septal patch.80 Surgical reconstruction with conservative techniques or prosthetic substitution of the valve has yielded relatively unsatisfactory results.81 Some patients present stenosis of the left ventricular outflow tract of subvalvular location. The obstruction can be anatomic and permanent, due to a membrane or subpulmonary ridge, or functional and dynamic, due to inversion of the curvature of the ventricular septum and the anterior systolic movement of the mitral valve.

Arterial correction

Arterial correction with the Jatene technique is replacing atrial correction in most hospital centers.82 It can be performed with a relatively low mortality in centers with experience and the intermediate-term results of the patients who survive are excellent.83,84 The most frequent complication is pulmonary supravalvular stenosis,85 followed by the less frequent aortic supravalvular stenosis. One reason for long-term concern is the disproportional expansion of the anatomic pulmonary root (aortic neoroot) that some patients have.86 This dilation can induce annulectasia and, in fact, the incidence of postoperative aortic
insufficiency is too high. However, the main reason for long-term concern after arterial switching is the state of coronary circulation. The transfer of the coronary arteries to the aortic neoroot is a delicate process that must be carried out with extreme care in order to avoid causing immediate catastrophes. However, a relatively high incidence of stenosis or occlusion of main coronary arteries has been detected even in patients who survive the intervention with no apparent perioperative complications. The prevalence of sudden death years after surgery is not unappreciable.

**SINGLE-VENTRICLE HEART**

In Table 5 is shown a list of the main CHDs that do not allow surgical reconstruction while maintaining a normal biventricular physiology. These are generally serious and complex CHDs with scant natural survival to adulthood and cardiologists for adults are not very familiar with them. The pediatric cardiologist sees them as a group of very heterogeneous problems that require an individualized diagnostic analysis and therapeutic approach. Cardiologists who treat adults must approach them as if they were a homogeneous disease. This way, all their anatomic complexity can be simplified by considering them as a single anatomic or functional ventricle. What matters most to cardiologists for adults is whether survival is spontaneous or previous interventions have been carried out in childhood.

**From Glenn to Fontan**

In 1958, Glenn established the therapeutic basis for eliminating the right side of the heart from the pulmonary circuit as an alternative to systemic-pulmonary fistulas. Glenn’s original intervention consisted of end-to-end anastomosis between the superior vena cava and right pulmonary artery. The technique did not have much initial acceptance due to fear that the venous flow was not sufficient to keep the pulmonary artery permeable, but the long-term clinical experience in many patients has been excellent. The Glenn operation acquired new interest when, in 1971, Fontan and Baudet described a palliative technique for the surgical treatment of tricuspid atresia. The Fontan technique completed the Glenn technique by carrying out a direct connection between the right atrium and pulmonary trunk or the rudimentary right ventricle.

It was soon observed that some of the pathophysiological hypotheses on which the Fontan technique rested were not correct and many technical aspects were modified. The first to disappear were the valvular prostheses at the inlet of the inferior vena cava and atrio pulmonary connection. The second concept to be revised was that bloodflow from the lower half of the body (60% of the total) went to the left lung, which is smaller (40% of capacity). This motivated a revision of the Glenn technique, maintaining the central connection of the two pulmonary arteries (bidirectional Glenn). The third concept to be revised was that of the distended right atrium as a hydraulic pump. In fact, the contractile function of the distended right atrium ceases at high pressures, which increases the reservoir effect, blood stasis, and atrial arrhythmias. These deleterious effects can be avoided by directly connecting the inferior vena cava to the pulmonary artery by means of an intra-atrial conduit (lateral tunnel) or a completely extracardiac conduit. The last concept to be revised was that of watertight compartmentation of the new cavopulmonary circuit. In order to avoid the adverse effects of a sharp rise in venous pressure, partial fenestration of the Fontan technique is used, leaving a transitory residual ASD that allows excess atrial pressure to be relieved in the immediate postoperative period.

**Expansion of the Fontan technique**

In initial experiences with the Fontan technique, the selection criteria for patients were very strict and are included in the famous decalogue of Choussat et al. In the 1980s, the selection criteria were expanded. The intervention was carried out not only in patients with tricuspid atresia, but also in those with different forms of single ventricle and other complex congenital heart diseases. Age limits were lowered, cardiac rhythm ceased to be a limiting factor, and anatomical and functional criteria became laxer. The result was a wave of moderate optimism which led to the operation of many patients with these techniques. In the 1990s, this optimism was replaced by a more realistic view. Fontan himself has clearly established that, even under the «perfect» conditions of a Fontan without sequelae, there is a progressive decrease in functional capacity and survival that cannot be attributed to anything except the circulatory status of the correction per se. Although longer-term results are still not

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**TABLE 5. Congenital heart diseases that preclude biventricular reconstruction**

<table>
<thead>
<tr>
<th>Condition</th>
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<tr>
<td>Anatomic single ventricle</td>
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<tr>
<td>Tricuspid atresia</td>
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<tr>
<td>Pulmonary atresia with hypoplastic right ventricle</td>
</tr>
<tr>
<td>Large or multiple ventricular septal defects</td>
</tr>
<tr>
<td>AV valve straddling</td>
</tr>
<tr>
<td>Mitral atresia or hypoplasia</td>
</tr>
<tr>
<td>AV canal with unbalanced ventricles</td>
</tr>
<tr>
<td>Extreme Ebstein anomaly</td>
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<tr>
<td>Syndrome of hypoplastic left heart</td>
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</table>
available, 5 to 15-year follow-up studies have demonstrated that the 10-year survival is only 60%. In this time, almost one-third of the survivors have had to be reoperated. 20% present cardiac arrhythmias that require antiarrhythmic medication or pacemakers, and 7-10% have hypoproteinemia due to protein-losing enteropathy. At 5 years of the operation, more than one-third have died or have a worse functional state than before surgery.102

Obstruction and thrombosis

In Table 6, the main complications of long-term Fontan circulation are described. The atrial or cavopulmonary connections may become obstructed by stenosis or kinking of the pulmonary arteries, retraction of suture lines, intrinsic degeneration of valvular prostheses, neointimal proliferation of conduits, calcification and the rigidity of prosthetic materials, or intraluminal thrombosis. The system created by Fontan surgery is very sensitive to stasis phenomena because venous flow is slow, and thromboembolic complications are common (Figure 7). The high incidence of thrombosis found in some series suggests that most adult patients should receive permanent anticoagulant treatment if no specific contraindications are present.103 Another long-term complication is the obstruction of the left ventricular outflow tract. Obstruction generally takes place in the VSD or bulboventricular foramen (Figure 8). The subaortic obstruction can be interpreted as a process of myocardial remodeling that occurs in response to changes in ventricular geometry induced by the reduction of the volumetric overload that transforms eccentric hypertrophy into concentric hypertrophy.104 An infrequent mechanical complication with very somber consequences is obstruction of the drainage of pulmonary veins, which can be partially occluded as a consequence of morphological changes in the right atrium after surgery.105

Persistent shunts and valvular insufficiency

After Fontan surgery, many patients have persistent shunts. These shunts can be produced by dehiscence of patches or lines of suture, neoformation of venous collaterals, distal aortopulmonary anastomoses, or the development of pulmonary arteriovenous fistulas. In some patients with an operated single ventricle, AV valve insufficiency can appear. The insufficiency can appear in very late phases of evolution and may have a progressive nature.106

Atrial arrhythmias and congestive heart failure

Atrial arrhythmias, especially atrial flutter and tachycardia due to macroreentry, are frequent after the Fontan procedure, associated with sudden death, and potentially refractory to all types of treatment, including drugs, antitachycardia pacing, and radiofrequency ablation.107 The prevalence of atrial arrhythmias in the follow-up of adults with Fontan physiology varies from 40% to 60% in different studies.108,109 Patients with atrial arrhythmia have a greater incidence of heart failure, atrial thrombosis, reoperation for conduit obstruction, AV valve incompetence, and ventricular dysfunction.110 Many patients with tricuspid atresia and single ventricle have ventricular dysfunction before surgery.111 Unfortunately, the Fontan intervention does not improve myocardial function in many cases, particularly when the operation is performed in patients over 10 years old.112

TABLE 6. Complications of the Fontan operation

<table>
<thead>
<tr>
<th>Anatomic complications</th>
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<tbody>
<tr>
<td>Obstruction or stenosis of connections</td>
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<tr>
<td>Atrial thrombosis and thromboembolic phenomena</td>
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<tr>
<td>Persistent or neoformed shunts</td>
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<tr>
<td>Pulmonary vein obstruction</td>
</tr>
<tr>
<td>AV valve insufficiency</td>
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<tr>
<td>Subaortic stenosis</td>
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<tr>
<td>Symptomatic arrhythmias</td>
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<tr>
<td>Atrial flutter or fibrillation</td>
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<tr>
<td>Incisinal tachycardia</td>
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<tr>
<td>Ventricular tachycardia</td>
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<tr>
<td>Ventricular dysfunction</td>
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<tr>
<td>Congestive heart failure</td>
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<tr>
<td>Protein-losing enteropathy</td>
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</tbody>
</table>

Fig. 7. Transesophageal echocardiogram in a patient with a single-ventricle heart corrected with the Fontan technique. The atrio pulmonary connection (large arrow) and a large mural thrombus in the wall of the right atrium (small arrows) are evident. RA indicates right atrium; LA: left atrium.
Protein-losing enteropathy

One of the worst long-term consequences of the Fontan operation is protein-losing enteropathy, which produces refractory ascites, generalized edema, and massive pleural effusion. The syndrome is infrequent in the first months or years after the operation, but its incidence increases with time. The clinical condition is similar to nephrotic syndrome and, like this condition, is produced by severe hypoalbuminemia. The cause of hypoalbuminemia is a chronic loss of proteins by the digestive system induced by the excessive and persistent increase in the venous pressure of the lower cava and portal system, with secondary lymphangiectasia. The diagnosis is based on the clinical condition, hypoalbuminemia, and the demonstration of increased elimination of α1-antitripsin in stool. The inhibitors of angiotensin-converting enzyme can be effective, but it is sometimes necessary to remove the Fontan connection or induce ASD to relieve central venous pressure. A chance observation has suggested that heparin treatment can be effective in improving hypoalbuminemia and the water-salt retention syndrome.

Revision of the Fontan operation

In patients with symptomatic atrial arrhythmias or severe protein-losing enteropathy that does not respond to medical treatment, revision of the Fontan intervention in adult life can be necessary. If stenosis exists in the connections, prosthetic materials, or pulmonary arteries, surgical reconstruction with enlargement of the obstructive areas may be sufficient. Patients with obstruction of the pulmonary veins or refractory atrial arrhythmias can benefit from complete atrial exclusion with an extracardiac Fontan or lateral tunnel. The subaortic obstruction can be corrected by muscular resection or a connection between the proximal pulmonary artery and ascending aorta. If the cause is myocardial dysfunction, the therapeutic alternatives are scant, but the clinical situation of some patients can be improved by undoing the Fontan modifications. Patients in an advanced functional class with no reasonable possibility for surgical reconstruction can still look to heart transplantation as an alternative solution.

NEEDS OF ADULTS WITH CONGENITAL HEART DISEASE

The large increment in the number of patients with CHD who will reach adulthood in the coming decades demands careful consideration of the new needs for care that are being generated, who will be responsible for providing care, and where it will be carried out. The physiology of many previously repaired or palliated CHDs can be difficult to understand for cardiologists without special training in pediatric cardiology. On the other hand, most of the medical problems of adults are beyond the training of pediatric cardiologists. The placement of these patients in wards, outpatient clinics, or examination rooms is also complicated. A pediatric setting is unsuitable for the continued care of older adults, but most cardiology departments are unprepared to handle such patients. The traditional barrier between pediatrics and general practice must disappear because teams are needed in which pediatric cardiologists and heart surgeons work hand in hand with cardiologists and heart surgeons specialized in adult patients. Adult congenital heart disease units (ACHDU) have been created in Europe and North America for this purpose. These units have been created to address a specific need, but it is now evident that their growth and development must be planned.

Each ACHDU should be formed by at least one cardiology specialist and an expert in CHDs, generally from pediatric cardiology, but heart surgeons expert in pediatric and adult problems must also participate, as
well as anesthetists with training and experience in both fields. Invasive (catheterization, electrophysiology) and non-invasive studies (echocardiography, stress techniques, dynamic electrocardiography, radionuclide medicine, magnetic resonance imaging) must be performed preferentially in adult units by specialists who are experts in CHDs. Hospitalization and outpatient care are best handled in hospitals for adults, but care by personnel who are experienced in the assessment and treatment of CHDs must be guaranteed. The ACHDU should also have access to a clinic for high risk pregnancies, a cardiac rehabilitation unit with special knowledge of congenital problems, a psychiatry or support psychology unit for adolescents, and a social services unit with special interest in the social and occupational problems of disabled young people. A fluid relation with the areas of clinical genetics (genetic counseling), hematology (blood dyscrasias), neurology (neurological syndromes), traumatology (skeletal malformations), and other specialized areas is important. The ACHDU must be structured in the setting of a tertiary teaching hospital and must have a close relation with one or more pediatric cardiology departments, which supply the patients and must be considered a natural and complementary extension. Ideally, each ACHDU should also be related with a heart transplantation unit with special experience in patients with CHDs. The American Heart Association has defined the need for ACHDU as one unit per 5-10 million inhabitants. For this reason, in Spain it would be necessary to plan for the development of no more than 4-8 units of this type.

Not all specialized care for adults with CHD can and should be provided in the ACHDU. In these units, only patients with complex heart diseases or serious complications should be followed up. The responsibility for most care inevitably lies with clinical cardiologists who have little training in CHDs. It has been estimated that 45% of adults with CHD do not need routine follow-up in a specialized unit. For another 30%, the responsibility for care must be shared between the specialized unit and clinical cardiologists. Ideally, each adult patient with CHD would have to be examined at least once in an ACHDU and returned to the community if a highly specialized follow-up is not necessary. It is important that there be direct communication between the extrahospital cardiologist and ACHDU, both for referral of patients and to consult on specific problems. Each clinical cardiologist should know the referral center. The referral center, with the support of institutions, should develop programs for continuing education to facilitate the work of cardiology specialists who have not had access to specific training. Institutions must also reconsider the training of ACHDU cardiology experts. Experts may come from the pediatric setting (pediatric cardiologists) or adult setting (clinical cardiologists), but in both cases they need additional training. The 23rd Conference of Bethesda has classified this additional training into three levels. Level 1 is acquired by means of accredited courses on the special problems of adults with CHD. It should be part of the training of all residents in cardiology and pediatric cardiology and extend to all clinical cardiologists in the community. Level 2 is acquired by a year of full-time post-residence in an ACHDU and level 3, by two years of continuous work in an ACHDU. Whether similar levels of training will be required in Spain is something that the Sociedad Española de Cardiología (Spanish Society of Cardiology) and healthcare and educational institutions will have to consider.

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