UP DATE

Clinical Decision Making Based on Cardiac Diagnostic Imaging Techniques (V)

Evaluation of Congenital Heart Disease in Adults

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Improvements in the diagnosis and surgical treatment of congenital heart disease during infancy and childhood have resulted in an outstanding increase in the prevalence of these entities during adulthood. Congenital heart disease in the adult represents a new diagnostic challenge to the consultant cardiologist, unfamiliar with the anatomical and functional complexities of cardiac malformations. Assessment of adult congenital heart disease with imaging techniques can be as accurate as in children. However, these techniques cannot substitute for a detailed clinical assessment. Physical examination, electrocardiography and chest x-rays remain the three main pillars of bedside diagnosis. Transthoracic echocardiography is undoubtedly the imaging technique which provides most information, and in many situations no additional studies are needed. Nevertheless, ultrasound imaging properties in adults are not as favorable as in children, and prior surgical procedures further impair image quality. Despite recent advances in ultrasound technologies such as harmonic or contrast imaging, other diagnostic procedures are sometimes required. Fortunately, transesophageal echocardiography and magnetic resonance imaging are easily performed in the adult, and do not require anaesthetic support, in contrast to pediatric patients. These techniques, together with nuclear cardiology and cardiac catheterization, complete the second tier of diagnostic techniques for congenital heart disease. To avoid unnecessary repetition of diagnostic procedures, the attending cardiologist should choose the sequence of diagnostic techniques carefully; although the information this yields is often redundant, it is also frequently complementary. This article aims to compare the diagnostic utility of different imaging techniques in adult patients with congenital heart disease, both with and without prior surgical repair.

Key words: *Adult congenital heart disease. Echocardiography. Transesophageal echocardiography. Magnetic resonance imaging. Nuclear cardiology. Cardiac catheterization.*

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Evaluación de las cardiopatías congénitas en el adulto

Los avances en el diagnóstico y tratamiento quirúrgico de las cardiopatías congénitas (CC) durante la edad pediátrica están provocando un incremento excepcional de su prevalencia durante la vida adulta. Las CC en el adulto representan un desafío diagnóstico nuevo para el cardiólogo clínico, poco familiarizado con la complejidad anatómica y funcional de las malformaciones cardíacas. La evaluación con técnicas de imagen de las CC del adulto puede llegar a ser tan precisa como en el niño, pero estas técnicas no pueden sustituir a una valoración clínica detallada. La exploración física, el ECG y la radiografía de tórax siguen siendo los tres pilares básicos en los que se sustenta el diagnóstico clínico. La ecocardiografía transtorácica es, sin duda, la técnica de imagen que aporta mayor información y, en muchos casos, suficiente. Pero los pacientes adultos no tienen tan buena ventana ultrasónica como los niños, y las intervenciones quirúrgicas previas añaden mayor deterioro de la imagen. Aunque las nuevas tecnologías ultrasónicas, como la imagen armónica o la ecocardiografía de contraste, tratan de solventar esta diferencia, con frecuencia es necesario recurrir a otros métodos diagnósticos. Afortunadamente, la ecocardiografía transesofágica y la resonancia magnética son más fáciles de realizar de forma rutinaria en el adulto que en el niño, por cuanto no precisan anestesia. Estas técnicas, junto con la cardiología nuclear y el cateterismo cardíaco, completan los métodos diagnósticos de segundo nivel disponibles actualmente. Para evitar exploraciones innecesarias, el cardiólogo clínico debe elegir bien la secuencia de técnicas diagnósticas, cuya información en muchos casos es redundante, pero en otros complementaria. Este artículo trata de comparar el valor diagnóstico de las diferentes técnicas de imagen en el adulto con CC con o sin intervención quirúrgica previa.

Palabras clave: *Cardiopatías congénitas del adulto. Ecocardiografía. Ecocardiografía transesofágica. Resonancia magnética. Cardiología nuclear. Cateterismo cardíaco.*

ABBREVIATIONS

AV: atrioventricular.
ASD: atrial septal defect.
VSD: ventricular septal defect.
TEE: transesophageal echocardiography.
TTE: transthoracic echocardiography.
MRI: magnetic resonance imaging.
TF: teratology of Fallot.
TGV: transporting of the great ressel.

INTRODUCTION

According to current estimates, 85% of children born with congenital heart disease survive to reach adulthood.[1] The spectrum of congenital heart diseases in adults is changing markedly, and diseases such as teratology of Fallot (TF), transposition of the great vessel (TGV), or the univentricular heart are emerging as practically new diseases because of therapeutic processes performed during infancy.[2] Almost all surviving patients present residual lesions, sequelae or complications that may progress during adulthood.[3] Congenital heart disease in adults is an intriguing diagnostic challenge for the cardiologist, who is often unfamiliar with the anatomical and functional complexity of heart malformations. Transthoracic echocardiography (TTE) is indisputably the imaging technique that provides most information about patients with congenital heart disease. With this technique, pediatric cardiologists have an advantage over general cardiologists because the conditions for ultrasound techniques in children are very favorable and the small size of the thorax allows very high frequency transdu-cers to be used. These favorable conditions do not last into adulthood and prior surgical procedures may further deteriorate the images. Although new ultrasound techniques such as harmonic imaging or contrast echocardiography are an improvement, it is often ne-cessary to resort to other diagnostic methods such as transesophageal echocardiography (TEE), magnetic resonance imaging (MRI), nuclear cardiography or angiocardiography. This article reviews the usefulness of imaging techniques in the evaluation of adults with congenital heart disease.

ATRIAL SITUS

Almost all adult patients with congenital heart disease present situs solitus and a few present situs inversus. It is therefore not normally necessary to establish the atrial situs because clinical examination, ECG and chest x-ray are more than sufficient to define the position of the heart. In some complex congenital heart conditions, the atrial situs is not so obvious but this information is essential for us to proceed. An anatomical diagnosis is impossible if we do not know the position of the heart. Essentially, atrial situs can be situs solitus, inversus or ambiguous. Atrial situs is often concordant with the positioning of the abdominal organs and the bronchia, and a careful examination of the chest x-ray may suffice. But this is not necessarily true in complex heart conditions. For example, the liver may have a central position or the bronchia may be arranged symmetrically, and the situs has to be defined by the atrial structure itself.

Normally, the inferior vena cava communicates with the right atrium, and the path of the vein can be followed readily by subcostal echocardiography. This technique also defines the position of abdominal organs. In situs solitus, the liver and inferior vena cava are located to the right of the abdominal aorta, and the stomach and the spleen to the left. In situs inversus, the arrangement is the mirror image of situs solitus. When the situs is ambiguous the liver, abdominal aorta and vena cava are centrally located, while the stomach can be positioned to the left or right and the spleen is normally either absent (asplenia) or there are multiple spleens (polysplenia). In such patients, the connection of the vena cava still indicates the right atrium, but in some complex patients, the inferior vena cava may be doubled or there may be an interruption in this vein before it enters the thorax, with circulation continuing through the azygos or hemiazygos system.

The most prominent morphological feature of the atria is the structure of the appendages. The right appendage forms a triangle with a broad base and a blunt point. The left appendage is much longer and more pointed, with the form of a gloved finger with a narrow base. In adults, the appendages cannot easily be defined by TTE, but their morphology can be visua-lized readily by TEE.[4] Therefore, we will have to resort to this technique in some exceptional cases in order to define the atrial situs (Figure 1). Magnetic resonance imaging is even more effective than conventional echocardiography or TEE for determining the thoracic or abdominal situs, and may be the technique of choice in adults with heterotaxia syndromes[5] (Figure 2).

BASIC CHAMBER SEQUENCE

Before analyzing anatomical and functional disorders of the heart in complex congenital heart conditions, it is essential to determine the internal structure of the heart. The basic chamber sequence can be determined by considering four points:[6]

1. Communication between atria and ventricles.
2. Communication between ventricles.
3. Connection between ventricles and arteries.

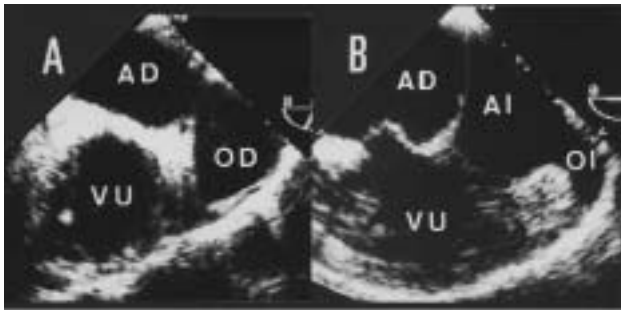

Fig. 1. Transesophageal echocardiogram in a patient with a single ventricle and juxtaposition of the appendages. Each atrium is identifiable by the morphology of the appendage. A: the right appendage (RA) is triangular with a blunt point. B: the left appendage (LA) is shaped like a gloved finger. RA indicates right atrium; LA, left atrium; CV, common ventricle.

4. The relationship between the aorta and the pulmonary artery.

Normally, the atrioventricular (AV) connection is concordant (the right atrium connects to the right ventricle and the left atrium to the left ventricle), the ventricles form a D-loop (right ventricle anterior and to the right). Connection of the ventricles to arteries is also concordant (the left ventricle is joined to the aorta and the right ventricle to the pulmonary artery) and the arterial circulation is normal (pulmonary valve anterior and to the left and the aortic valve posterior and to the right). Complex congenital heart conditions may, in contrast, have a very different blood flow. The AV connection may be concordant, discordant, ambiguous (indeterminate), have double outflow or not be connected. The ventricles may be arranged as a D-loop, an L-loop or be indeterminate (common ventricle). The arterial connection to the ventricles may be concordant, discordant, or there may be a single or double arterial outlet from a ventricle. The arterial arrangement can be normal in situs solitus, normal in situs inversus, or have a D- or L-transposition.

Often, the type of AV connection, possible presence of ventricular septal defect (VSD) and the type of arterioventricular connection can be accurately determined by TTE with apical view. The arrangement of the great arteries is easier to determine with transversal planes along the parasternal or subcostal view. In a few adult patients with complex congenital heart disease, it may be impossible to define the direction of blood flow. Multiplanar TEE provides very precise information in such patients on the type and mode of AV and ventri-cular-arterial connection, and on the morphology of each ventricle[7] (Figure 3).

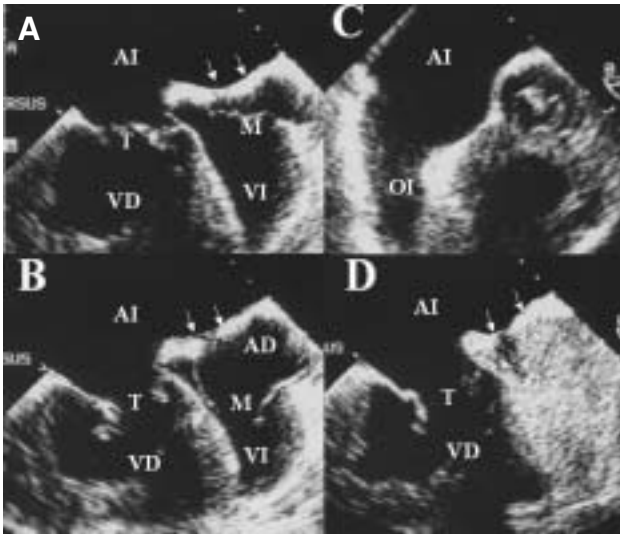

Fig. 3. Transesophageal echocardiogram of a patient with situs inversus and atrioventricular discordance. A and B (four-chamber plane in systole and diastole): the atrial septum (arrows) forms nearly a right angle with the ventricular septum; the right ventricle (RV), located to the right, is recognized by the morphology of the tricuspid valve (T), and the left ventricle (LV), located to the left, by the morphology of the mitral valve (M). C (transverse plane): the left atrium (LA), located to the right, is recognized by the morphology of the appendage (LApp). D: after injection of contrast, the right atrium (RA) and the left ventricle are filled, but there is no flow of contrast to the chambers of the right side.

VENOUS DRAINAGE ANOMALIES

The most frequent venous drainage anomalies in adults affect the venae cavae and do not have any functional repercussions, but an incidental anomaly

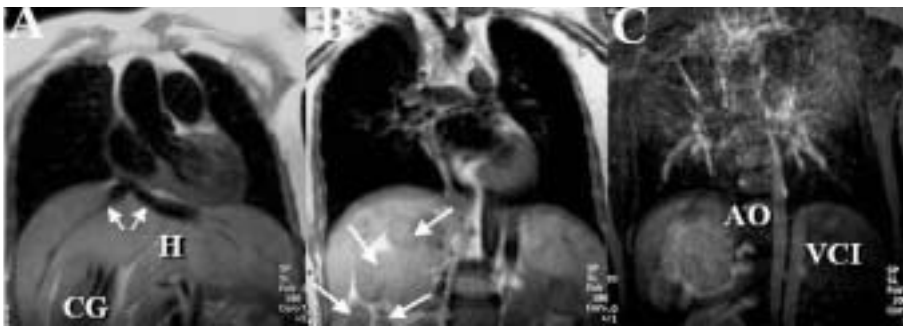

Fig. 2. Magnetic resonance imaging in a patient with left isomerism. A and B: SE T2 morphological images showing the «pancake» morphology of the liver in the central position (L), the gastric chamber (GC) to the right and the independent drainage of the suprahepatic veins (short arrows) that correspond to multiple spleens (polysplenia). C: magnetic resonance angiography showing the inferior vena cava (IVC) to the left of the abdominal aorta (AO).

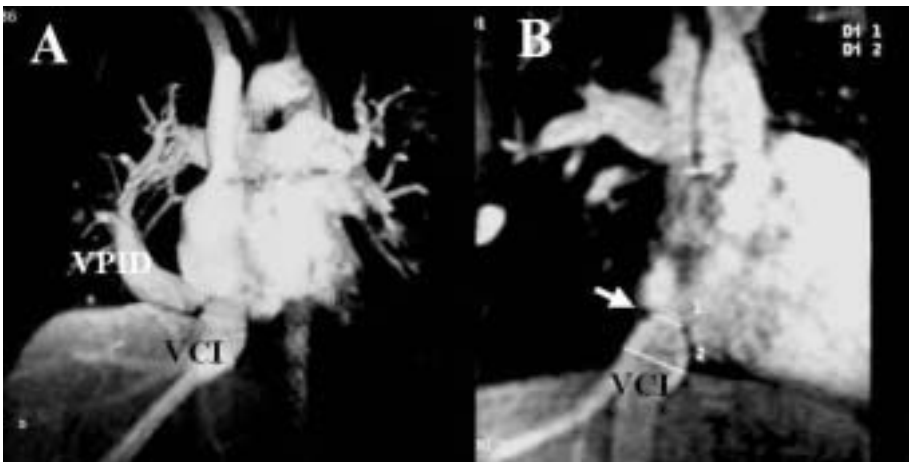

Fig. 4. Magnetic resonance imaging in a patient with scimitar syndrome. The right inferior pulmonary vein (RIPV) follows an anomalous course and joins the inferior vena cava (IVC). Stenosis of the IVC (arrow) occurred after surgical correction. The stenosis was resolved by angioplasty.

may confuse the interpretation of the heart images, whatever the technique used, or alter the surgical approach or invasive procedures. The most common anomalies are persistent left superior vena cava drainage to the coronary sinus and stenosis of the inferior vena cava with continuation via the azygos or homozygous system. Transthoracic echocardiography can detect these anomalies. The suprasternal view will show the presence of a superior left vena cava and determine whether the innominate vein connects the two superior venae cavae. We can inject sonicated serum into a peripheral vein to help delineate the drainage chamber of the different venous territories. However, echocardiography does not allow the entire anomalous venous course to be determined, and angiocardiography and MRI are needed for accurate diagnosis. Currently, MRI is the technique of choice for defining venous drainage anomalies in adult patients[8] because it can accurately delineate incidental anomalies, as well as the rare cases of non-incidental vena cava anomalies, such as drainage of the inferior vena cava or the left or right superior vena cava into the left atrium.

Magnetic resonance imaging is even more useful in the evaluation of pulmonary venous drainage. The most frequent anomaly in adults is partial anomalous drainage of pulmonary veins. Anomalous drainage occurs more frequently in the superior pulmonary vein, but it can also affect the left pulmonary veins or inferior pulmonary veins, particularly the right inferior pulmonary vein. An anomalous right superior pulmonary vein will normally drain into the superior vena cava or the junction between the superior vena cava and the right atrium, and it is associated with sinovenous atrial septal defect (ASD). Anomalous drainage of a superior left pulmonary vein normally occurs via a left vertical vein that runs into the innominate vein. Anomalous drainage of an inferior pulmonary vein (normally the right one) is usually to the inferior vena cava, giving rise to scimitar syndrome (Figure 4). Although these anomalous shunts may be much better assessed by TEE, the current technique of choice for diagnosis of these anomalies is MRI.[9]

ATRIAL SEPTAL DEFECTS

Interatrial shunts normally result from ASD within the confines of the fossa ovalis (ostium secundum). In 15% of patients, the defect is part of an anomalous development of the endocardial cushions, affecting the atrial septum immediately adjacent to the AV valve plane (ostium primum). Less than 10% of have a posterior defect, outside the confines of the fossa ovalis (sinovenous), which is usually associated with an anomaly in the venous drainage of the superior right pulmonary vein. The ASD may also rarely be located in the coronary sinus (unroofed coronary sinus) or affect the entire atrial septum (single atrium). Most patients with ASDs can be evaluated initially with TTE. The diagnostic sensitivity is very high for any type of ASD, though a sinovenous ASD sometimes causes difficulties for transthoracic examination, particularly if the subcostal view is inadequate. Transesophageal echocardiography is better than conventional echocardiography for determining the size, number, morphology and location of defects.[10] Transesophageal echocardiography is indicated:

1. In all patients suspected to have or with confirmation of sinovenous ASD, as the shunt is normally associated with vena cava drainage anomalies (overriding) and anomalous pulmonary veins that cannot be analyzed satisfactorily by TTE[11] (Figure 5).

2. Whenever the therapeutic approach requires accurate determination of the size, number, morphology or exact position of the defects.[10]

3. To determine the borders of the defect whenever percutaneous closure of the ASD is considered, and to predict the possibility of success with this technique[12]

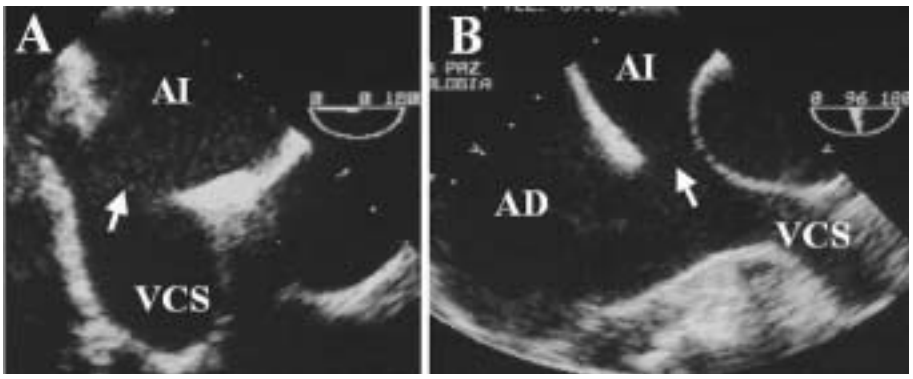

Fig. 5. Transesophageal echocardiogram in a patient with sinovenous atrial septal defect. A (transversal plane): the defect (arrow) is located between the superior vena cava (SVC) and the left atrium (LA). B (longitudinal plane): the SVC straddles both atria. RA indicates right atrium.

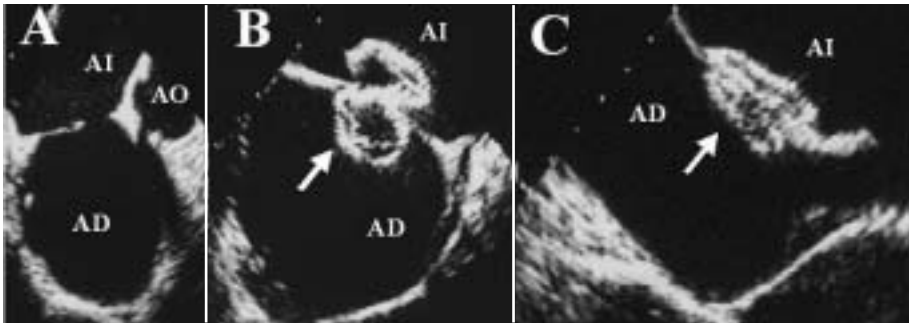

Fig. 6. A, B and C: transesophageal echocardiogram during placement of Amplatzer device (arrow) for percutaneous closure of an ostium secundum atrial septal defect. RA indicates right atrium; LA, left atrium.

(Figure 6).

The remaining patients can be managed using the findings from TTE, though we normally complete the information from this technique by performing radionuclide ventriculography to quantify the left-to-right shunt (Qp:Qs). The Qp: Qs ratio can also be eva-luated by conventional Doppler echocardiography, but use of an additional technique is advisable.[13] Cardiac catheterization should only be used in those patients with severe pulmonary hypertension because it is essential for calculation of the pulmonary vascular resistance and the response of pulmonary pressure to vasodilators.

Experience in recent years has shown that the persistence of a patent foramen ovale may frequently cause cryptogenic brain infarction in young patients, particularly if associated with aneurysm of the fossa ovalis.[14] Transesophageal echocardiography is the technique most used for diagnosis of patent foramen ovale, but we believe that detection is just as sensitive using contrast echocardiography with sonicated serum in conjunction with harmonic imaging. We currently only perform TEE when percutaneous closure of the foramen ovale is considered as a therapeutic option.

INTRA-ATRIAL MEMBRANES AND BAFFLES

Cor triatriatum is an uncommon heart disorder in which a membrane divides the left atrium into two chambers. The proximal chamber receives drainage from the four pulmonary veins and the distal chamber incorporates the left appendage and the atrial septum. When presence of this membrane forms a restrictive orifice joining the divided atrium, the clinical signs are similar to those of mitral stenosis but diagnosis is much harder using TTE. In contrast, diagnosis and assessment by TEE[15] are much easier. Thus this technique is indicated whenever this anomaly is suspected. Intra-atrial membranes in the right atrium are more common, though they are often incidental findings. They may take the form of a Chiari network, prolapse of venous valves or cor triatriatum dexter. Evaluation is adequate with TTE and it is not often necessary to resort to other techniques.

Intra-atrial baffles are introduced by heart surgeons to divide the atria and redirect atrial flow. Their main application is the atrial physiological correction of transposition of the great vessels. Two surgical techniques are commonly used. The Mustard technique uses only prosthesis material whereas the Senning technique uses the atrial wall itself and the septum to achieve atrial division. The clinical results and the echocardiographic image obtained afterwards are similar for both techniques. The resulting septation forms a pulmonary-venous canal that directs the flow of the pulmonary veins towards the tricuspid valve and a systemic venous canal that incorporates the axis of the cavae and directs the flow of the venae cavae towards the mitral valve. The echocardiographic evaluation is satisfactory in most cases but when dehiscence or ste-

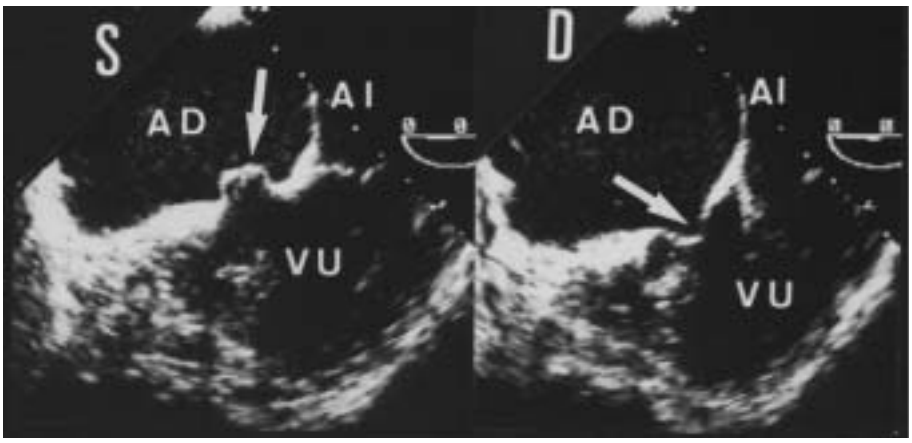

Fig. 7. Transesophageal echocardiogram during systole (S) and diastole (D) in a patient with a single ventricle and hypoplasia of the tricuspid valve (TV): the minimal diastolic opening of the valve (arrow) was not visible in the transthoracic echocardiogram. RA indicates right atrium; LA, left atrium; CV, common ventricle.

nosis of the baffles is suspected, it is necessary to resort to other techniques. Dehiscence or stenosis of baffles normally occurs at the entrance of the superior vena cava and can be satisfactorily visualized by TEE,[16] but when surgical repair or therapeutic intervention is considered (balloon and/or stent dilatation), it is necessary to perform a prior angiography.

ATRIOVENTRICULAR VALVE MALFORMATION

There is a broad range of AV valve malformations. There may be two well differentiated valves or a common AV valve. One valve may be missing, perforated, hypoplastic, stenotic or insufficient. The valve may override the septum with a sub-valvular apparatus in a single ventricle or in both ventricles. In adults, malformation of papillary muscles of the mitral valve is the most common finding. It is associated with aortic coarctation and is normally incidental. An acute malformation may behave like a single papillary muscle causing mitral stenosis, often associated with the Shones complex. Rarely, mitral stenosis in adults can be caused by the supravalvular mitral ring or congenital fusion of the commissures.

The most common cause of congenital mitral regurgitation is an isolated anterior cleft of the mitral valve. In canal-like malformations, mitral regurgitation is caused by a similar but anatomically different malformation in which tendinous cords between the cleft and the septum cause sphincter-like function. A double mitral orifice is an uncommon malformation that also leads to valvular dysfunction.

The most common malformation of the tricuspid valve is the Ebstein anomaly. Congenital stenosis of the tricuspid valve is a rare finding, but it may appear after septation of the common AV canal. Uncorrected tricuspid atresia is an uncommon heart condition in adults, but there are increasing numbers of patients with tricuspid atresia corrected using the Fontan procedure.

Generally, TTE with color Doppler and continuous-wave Doppler imaging allows an anatomical and functional assessment of the malformations of the AV valves. When this technique is not entirely satisfactory, TEE allows a more precise assessment of these malformations[17] (Figure 7). Transesophageal echocardiography is particularly useful for intraoperative assessment when valve repairment techniques are performed.[18] Transesophageal echocardiography is also useful to evaluate patients with Ebstein anomaly awaiting surgery to determine whether valve repair might be successful. Multiple adherences of the tricuspid valve to the anterior ventricular wall hinder suitable surgical repair (Figure 8).[19]

When assessing AV valve malformations, cardiac catheterization and angiocardiography are only indicated in exceptional circumstances.

FONTAN CIRCULATION

There are many congenital heart diseases that cannot be surgically reconstructed while still retaining the function of both ventricles. These are patients with a single anatomical or functional ventricle, including tricuspid or mitral valve atresia, an AV valve overriding tensor apparatus on either side of the septum, pulmonary or aortic atresia with hypoplasia of the underlying ventricle, complex congenital heart conditions with unbalanced ventricles, or extreme forms of the Ebstein anomaly. In 1958, Glenn laid the foundations for partial bypass of the right side of the heart as a therapeutic alternative, suturing the end of the superior vena cava directly to the end of the right pulmonary artery (classic Glenn anastomosis) or performing an end-to-side shunt (bidirectional Glenn anastomosis). In 1971, Fontan reassessed the Glenn shunt and performed the first complete bypass of the right ventricle, connecting the right atrium to the pulmonary artery. During the eighties and nineties, the Fontan procedure underwent many changes and, currently, most surgeons prefer direct anastomosis of both venae cavae to the pulmonary

artery (total cavopulmonary connection) using a lateral tunnel in the right atrium or a completely extracardiac conduit to extend the inferior vena cava.

Patients with Fontan circulation who reach adulthood are exposed to numerous complications: stenosis or narrowing of the connections or pulmonary arteries; thrombosis of the right atrium or Fontan connection; persistent or newly formed shunts; stenosis of pulmonary veins caused by the large increase in size of the right atrium; insufficiency of the AV valves; ventricular dysfunction or subaortic stenosis through partial closure of the bulboventricular foramen.[2] Monitoring of these patients with TTE provides very limited anatomical and functional information. The information provided by TEE for detecting atrial thrombosis and stenosis of the Fontan connection or pulmonary veins is much more useful[20,21] (Figure 9). Magnetic resonance imaging may be indicated to assess the morphology of the main pulmonary arteries and the connection between the superior cava and the pulmonary artery.[22,23] Both techniques are complementary and should be carried out while monitoring patients with Fontan circulation when complications are suspected. Heart catheterization is indicated for assessing the pressure and resistance of the pulmonary arteries before performing any further intervention.

VENTRICULAR SEPTAL DEFECTS

The most common congenital heart disease in newborn children is VSD but, fortunately, the defect usually closes spontaneously during the first years of life. Patients who reach adulthood with a VSD usually have a restrictive defect, with little left-to-right shunt and normal pulmonary pressure. In most cases, the VSD will eventually close during adulthood. Occasionally, a patient with a large VSD and pulmonary hypertension reaches adulthood with Eisenmenger syndrome, but this is currently the exception. Some patients with restrictive VSD can develop progressive subinfundibular stenosis of the right ventricle (double chamber right ventricle) as adults. Others develop aortic valve regurgitation, fixed subaortic stenosis or tricuspid regurgitation. Infectious endocarditis is a relatively frequent complication that may affect the mural endocardium, the aortic valve or the tricuspid valve.

All patients with VSD should be monitored with clinical examinations and echocardiographs while the shunt remains open. Transthoracic echocardiography with color Doppler imaging is an excellent technique for determining the anatomical position of the VSD (perimembranous, trabecular, inlet septal, subarterial or ventriculoatrial), the size of the defect, the magnitude of the shunt and the systolic pressure of the right ventricle and pulmonary artery. This technique is also very useful for early detection of possible complications. Transesophageal echocardiography and MRI are used only in patients with a very poor transthoracic view or when conventional echocardiography does not provide detailed information on the complications (aortic or tricuspid regurgitation mechanism, suspicion of endocarditis not shown by TTE). Nuclear angiocardiography should be performed In patients with moderate left-to-right shunt to quantify the Qp:Qs ratio.

Cardiac catheterization is indicated in patients with high pulmonary blood pressure for determination of the vascular pulmonary resistance and response to vasodilators. Residual shunts are sometimes detected with Doppler color imaging in patients who have previously undergone an intraventricular patch operation. The shunt is usually restrictive, but it may increase over time. The patch may retract and become calcified making anatomical assessment of the defect difficult, but Doppler color imaging is not affected. When the

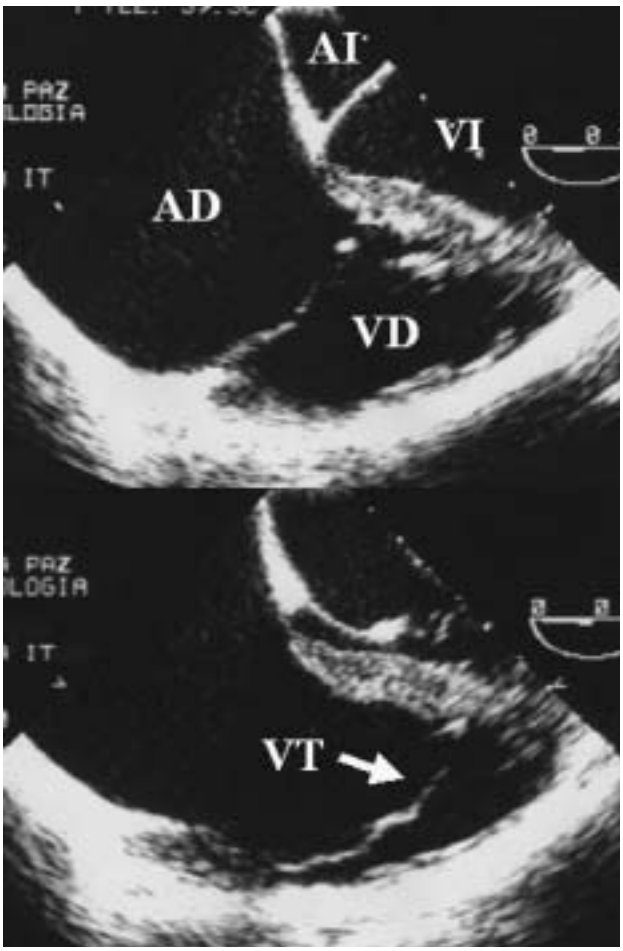

Fig. 8. Transesophageal echocardiogram of a patient with Ebstein anomaly of the tricuspid valve (TV). The septal valve is displaced towards the apex. The anterior valve is in its normal position and is large and hypermobile, with no adherence to the ventricular wall, allowing plastic reconstruction of the valve. RA indicates right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle.

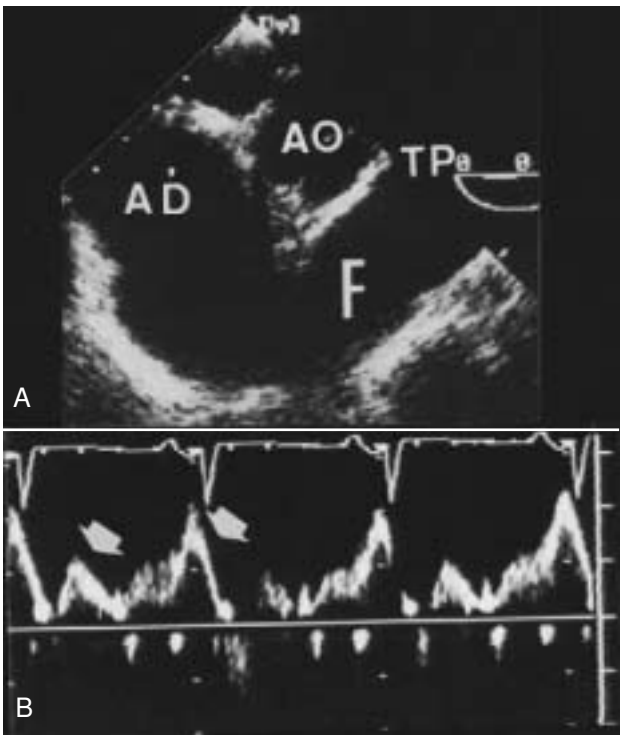

Fig. 9. Transesophageal echocardiogram in a patient with Fontan circulation. A: two-dimensional image revealing the atriopulmonary connection (F). B: pulsed Doppler image of atriopulmonary flow with two diastolic waves (short arrows). RA indicates right atrium, AO: aorta; PT: pulmonary trunk.

patch is large, the direction of the shunt may be extremely diverse (interventricular, ventriculoatrial or aortoventricular); nevertheless, most cases can be assessed by TTE.

VENTRICULAR MORPHOLOGY AND FUNCTION

In congenital heart diseases, as in acquired heart disease, ventricular function is the main prognostic factor, but in congenital heart diseases, pulmonary ventricular function is as important as systemic ventricular function. Moreover, the systemic ventricle may have left, right or indeterminate morphology, which makes assessment of ventricular function more difficult. The left ventricle is recognized by its smooth-wall structure and the right by its trabecular structure, but the main defining feature is the morphology of the AV valve. When there are two ventricles, the right one is always connected to the tricuspid valve and the left to the mitral valve. When a double-inlet ventricle is present, the morphology is left if it has an outlet chamber and right if both arterial vessels leave the main ventricle. Echocardiographic assessment of left ventricular function is the same in congenital heart disease and acquired heart disease, but echocardiographic assessment of right (pulmonary or systemic) ventricular function in congenital heart disease is much more difficult. The complicated structure of the right ventricle does not allow direct quantitative determination of the ejection fraction, though subjective visual estimation of systolic function is just as valid. Parameters that measure the diastolic diameter of the inflow tract, systolic shortening of the area in the plane of the four cardiac chambers, the global ventricular function (Tei index) or total internal diameter of the tricuspid ring are all useful for quantifying right ventricular function. However other techniques are often necessary to determine the ejection fraction and ventricular volumes. Equilibrium radionuclide ventriculography is a method that has been shown to be of great use for determining the volumes and function of the right ventricle,[24] though the most reliable method currently in use is MRI.[25,26] This technique allows the morphology and structure of both ventricles to be determined and the degree of hypertrophy or dilatation of the cavity and overall systolic function to be assessed with greater precision, regardless of the ventricular morphology.

OUTFLOW TRACT OF THE LEFT VENTRICLE

Obstruction of the outflow tract of the left ventricle may be valvular, subvalvular or supravalvular. Transthoracic echocardiography can usually suffice for locating the obstruction and determining its morphology, and continuous Doppler imaging allows the maximum instantaneous gradient and the mean gradient at the obstruction to be determined. Occasionally, patients with fixed subaortic stenosis with marked septal hypertrophy require TEE to differentiate localized subaortic stenosis from obstructive hypertrophic myocardiopathy[27] and, in some patients with aortic supravalvular stenosis, MRI[28] allows the morphology of the obstruction and the ascending aorta to be determined.

Aortic valve regurgitation is also assessed by conventional echocardiography with color Doppler imaging. Nevertheless, in patients who are to undergo surgery preserving the valve, TEE provides more information on the functional anatomy of the aortic root and the regurgitation mechanism (dilatation of the sinotubular junction, asymmetry of the sinuses, valvular prolapse and tenting, leaflet retraction or perforation).[29] As in the case of mitral insufficiency, this technique is also very useful for intraoperative monitoring of the aortic valve repair.

OUTFLOW TRACT OF THE RIGHT VENTRICLE

As in the left ventricle, obstruction of the right outflow tract may have a valvular, subvalvular or supravalvular position. Normally, pulmonary valve stenosis can be accurately assessed with TTE and continuous Doppler imaging, though the assessment of subvalvu-

lar or supravalvular obstruction may be more difficult. Subvalvular obstruction may affect the infundibulum (excessive hypertrophy or infundibular hypoplasia) or be subinfundibular. This latter case appears occasionally in adults with VSD because hy-pertrophy of the septomarginal bands can cause ventricular obstruction, dividing the right ventricle into two chambers (double-chambered right ventricle). Parasternal echocardiogarphic assessment is difficult because the anomalous band can be confused with the normal moderator band, the jet from the obstruction is confused with that of the VSD, the pressure gradient is underestimated (because the direction of the jet is transversal to the ultrasonic plane) and the increase in systolic pressure in the right ventricle is attributed to pulmonary hypertension.[30] Subcostal echocardiography is much more appropriate for locating the obstruction and assessing the ventricular gradient, but subcostal planes are not always possible in adults. Transesophageal echocardiography can determine the morphology of the subinfundibular obstruction more precisely, and it is also useful for intraoperative monitoring of resection of the anomalous muscle band.[31]

Supravalvular pulmonary stenosis can occur in the pulmonary main artery or in peripheral branches. Stenosis of the pulmonary artery caused by a fibrous ring is uncommon, but it has to be differentiated from valvular stenosis. Transthoracic echocardiography is normally satisfactory, though the images provided by TEE or MRI are clearer. The most common cause of stenosis of the main pulmonary artery in adult patients is the postoperative sequela known as pulmonary banding. Despite resection of the cerclage during definitive repair of the heart disorder, some patients still have supravalvular pulmonary stenosis, which can be adequately assessed with TTE. Another very distinct case is supravalvular stenosis located downstream from the pulmonary bifurcation. This malformation may occur as an isolated anomaly, but more often it may be caused by a residue, sequela or complication in patients who have undergone surgery for TF or pulmonary atresia with VSD. Although suprasternal echocardiography or TEE may help to determine the morphology of pulmonary branches, the technique of choice is MRI. Gadolinium-enhanced three-dimensional pulmonary angioresonance reconstruction offers an excellent image of the central pulmonary vascular tree[32] (Figure 10).

Adult patients who have undergone surgery for congenital heart disease may have stenosis of the outflow tract of the right ventricle due to the intrinsic degene-ration of a biological pulmonary prosthesis. Pulmonary prostheses can be assessed by TTE, but prosthetic conduits can be much more difficult to determine using this technique. Often, the conduits between the right ventricle and the pulmonary artery are located behind the sternum in the upper region of the thorax, and this makes study of these conduits by echocardiography more difficult. We have found that TEE is normally very effective for examining prosthetic conduits and for determining the pressure gradient, but when these conduits become highly calcified, the intraprosthetic ultrasound image is poor. Magnetic resonance imaging may overcome these difficulties and help to determine the reason for the obstruction and its position,[33] though cardiac catheterization and angiocardiography are often required.

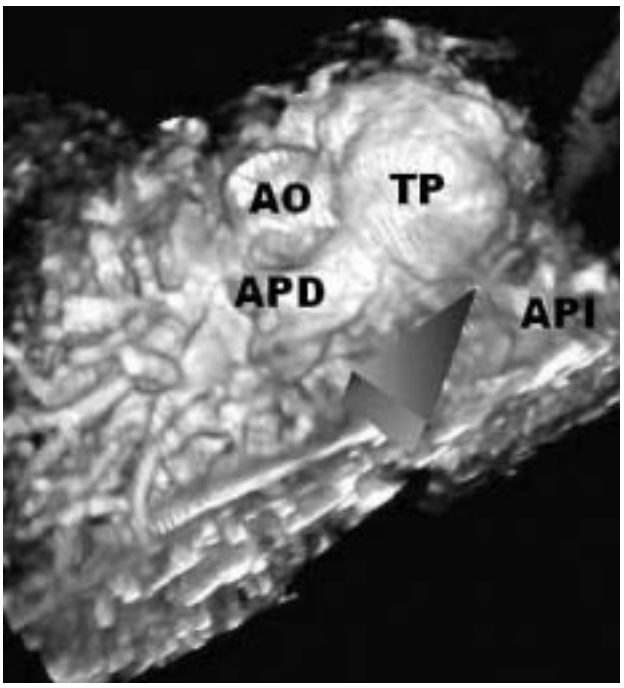

Fig. 10. Magnetic resonance image of a patient teratology of Fallot corrected with a transannular patch. MR angiograph sequence in which filiform stenosis is seen at the origin of the left pulmonary artery (arrow head). AO indicates aorta; RPA, right pulmonary artery; LPA, left pulmonary artery; PT, pulmonary trunk.

Pulmonary valve regurgitation is the most common valve disorder in adult patients with congenital heart disease. This insufficiency is rarely itself congenital, but it almost always appears as a postoperative sequela in patients with critical pulmonary stenosis or TF corrected by a transannular patch enlargement. Mild pulmonary valve regurgitation after this type of surgery is usually well tolerated for years but, in the long term, it causes dilatation and systolic dysfunction of the right ventricle, secondary tricuspid insufficiency, right heart failure and sustained ventricular or atrial arrthymias. Pulmonary valve insufficiency may be assessed either by conventional echocardiography with color Doppler and pulsed Doppler imaging. Particular attention must be given to the assessment of the systolic and diastolic function of the right ventricle, and the aforementioned limitations of the echocardiography should be recognized.

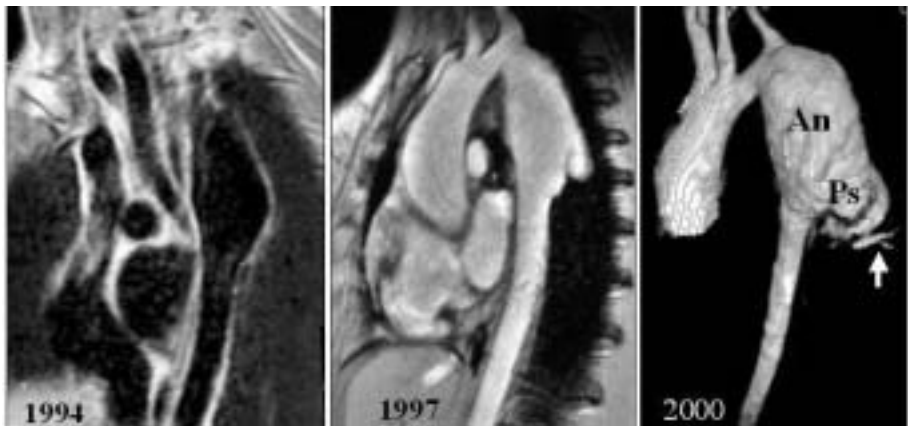

Fig. 11. Monitoring of a patient operated on for aortic coarctation by aortoplasty with Dacron graft. In 1994 (left) aneurysmal dilatation of the aortoplasty was present. In 1997 (middle), a pseudoaneurysm was seen at the distal end of the graft. In 2000, partial thrombosis of the pseudoaneurysm was found which had perforated a subsegmental bronchus (arrow), provoking intense hemoptysis. An indicates aneurysm; Ps, pseudoaneurysm.

PATHOLOGY OF THE THORACIC AORTA

The congenital heart condition that most often affects the thoracic aorta is coarctation. At present, most patients with aortic coarctation are diagnosed and treated during infancy, but the defect may not be detected until adulthood. The diagnosis of coarctation in adults is based on clinical findings. Suprasternal echocardiography allows the flow characteristics through the coarcted area to be determined and the gradient to be estimated, but the morphology of the coarctation is analyzed much better by MRI. This technique provides very precise information on the anatomy of the aortic isthmus, the minimum intraluminal diameter and flow along the axis of the coarctation.[34] Aortography is only required when a therapeutic procedure involves a dilatation balloon or stent. Adult patients who underwent surgical repair of coarctation during infancy may present aortic complications such as re-coarctation or formation of aneurysms. In such cases, MRI is also the technique that provides most diagnostic benefit (Figure 11).[35] Arterial rings are uncommon malformations in the adult, and are usually incidental anomalies. Magnetic resonance imaging is also the technique of choice for defining the anomalies of supraaortic trunks and arterial rings.[36]

Aneurysmal dilatation of the ascending aorta is a common complication in patients with Marfan syndrome, though it also appears frequently in patients with coarctation (whether they have undergone surgery or not), a bicuspid aortic valve and TF, or pulmonary atresia with VSD. Dilatation of the ascending aorta in these cases is caused by disease of the middle aortic layer and may be the cause of aortic regurgitation, dissection, intramural hematoma or wall rupture. Most authors recommend surgery when the inner lumen diameter exceeds 50 mm. Follow-up can be performed by echocardiography, but MRI is more reliable and is therefore considered the technique of choice.[37]

Aneurysms of sinuses of Valsalva are saccular invaginations shaped like a glove finger. They are not usually noticed until they cause a perforation of the right ventricle or atrium leading to a large left-to-right shunt and congestive heart failure. Diagnosis is possible by TTE, but differential diagnosis with VSD and aortic regurgitation is difficult. Transesophageal echocardiography provides a more exact definition of the morphology, position and drainage chamber, and can differentiate from a coexisting VSD if present. Therefore, TEE can be considered the technique of choice for the preoperative assessment of aneurysms of sinuses of Valsalva.[38]

AORTOPULMONARY FISTULAS

Persistence of patent ductus arteriosus causes a congenital aortopulmonary fistula readily detected by TTE with color Doppler imaging. Currently, this congenital heart disease is uncommon in adults. Using this technique, patent ducts are occasionally found in adults with no repercussions or continuous murmur. These incidental findings show the great sensitivity of color Doppler imaging for detecting this anomaly. At the other extreme, some patients with a large ductus arteriosus may develop pulmonary hypertension, which becomes clinically manifest in adulthood. Diagnosis by TTE in these cases is more difficult, and a TEE may be indicated (Figure 12).[39]

The most common aortopulmonary fistulas in adults with congenital heart disease are those provoked by the surgeon to increase pulmonary flow in cyanotic heart disease with pulmonary stenosis. Fistulas resulting from surgery are very diverse. The most common is anastomosis of the subclavian artery to the ipsilateral pulmonary artery (Blalock-Taussig shunt) or anastomosis of the ascending aorta to the right pulmonary artery (Waterston shunt), but increasing numbers of surgeons are using conduits of synthetic material (Gore-Tex) to connect systemic and pulmonary circulation. Surgical

fistulas are difficult to analyze using TTE or TEE, and so, in case of doubt about their patency, morphology, flow or complications, it is necessary to resort to MRI[40] or angiocardiography.

Systemic collaterals between the descending aorta and the pulmonary vascular tree are common in patients with pulmonary atresia and non-restrictive VSD (pseudotruncus). This may cause heart failure in adults because of a large left-to-right shunt, which can be corrected with unifocalization of pulmonary blood supply. The localization of systemic collaterals requires angiography and selective sounding of each collateral. Magnetic resonance imaging may be of great help in the localization of different collaterals before catheterization and prevent important collaterals from being missed during angiographic assessment.[32]

Some adult patients may present bronchopulmonary or pulmonary arteriovenous fistulas that are difficult to diagnose and locate. The diagnosis of these anomalies requires selective angiography, but MRI may be very useful for localization of the fistulas. Contrast echocardiography with sonicated serum is very sensitive for the detection of pulmonary arteriovenous fistulas, though the technique cannot locate them.[41]

CORONARY ARTERY ANOMALIES

There are many types anomaly of congenital coronary arteries. They are usually simple variations in the origin of the main branches, the most common being the circumflex artery starting from the right coronary artery or from the right coronary sinus as an independent trunk. The anterior descending artery originating from the right coronary sinus is less common but there may be important surgical implications when this defect is associated with TF. Occasionally, there is a single coronary ostium that gives rise to the three branches. The path of the branches is of more importance than their point of origin because exercise angina or sudden death during exercise may occur if the aberrant artery passes between the aortic root and the outflow tract of the right ventricle. Different congenital heart diseases (TF, complete or corrected TGV, double right ventricle outlet or truncus arteriosus) have a different coronary anatomy that must be determined before surgery. Selective aorto-graphy or coronary arteriography should always be performed in adult patients with complex congenital heart diseases before surgery to avoid damage to the left main artery during surgery. The major coronary artery anomalies are the origin of the left coronary trunk of the pulmonary artery, coronary fistulae and coronary aneurysms. Although these anomalies may be diagnosed by echocardiography (TTE or TEE) and by MRI, definitive diagnosis should be established by selective coronary arteriography.[42]

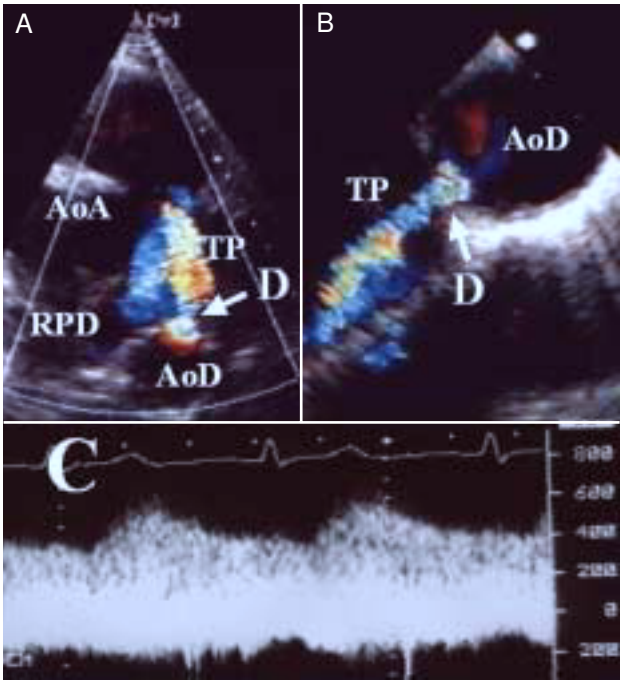

Fig. 12. Color Doppler transthoracic (A) and transesophageal (B) echocardiogram and continuous Doppler image (C) of a patient with patent ductus arteriosus. PT indicates main pulmonary artery; DAO, descending aorta; AAO, ascending aorta; RPB, right pulmonary branch; D, ductus.

FIRST CHOICE DIAGNOSIS PROCEDURES

Evaluation of imaging techniques for congenital heart diseases in adults can achieve the precision seen in children, but these techniques cannot replace a detailed clinical assessment. The clinical history should accurately record all previous interventions, symptoms and the functional state of the patient. The physical examination, ECG and chest x-ray are still the three basic pillars that support clinical diagnosis. Imaging techniques are complementary and should be guided by the clinical assessment. Transthoracic echocardiography with pulsed, continuous-wave and color Doppler imaging is the main imaging technique and this technique alone will often suffice. Transthoracic echocardiography can usually determine the atrial situs, the basic chamber sequence, defects in the atrial or ventricular septum, the patency of ductus arteriosus, the morphology and function of the AV valves and the outflow tract of the two ventricles, the ventricular structure and the quantitative function of the left or right ventricle and systolic blood pressure in the pulmonary artery, and to assess shunts and valvular regurgitations.

Transesophageal echocardiography, MRI, nuclear cardiology and cardiac catheterization are second level diagnostic techniques that are only indicated

TABLE 1. **Diagnostic procedures in adult patients with congenital heart disease**

	TTE	TEE	MRI	Isotopes	Catheterization
Atrial situs	+++	++	++	-	-
Heart structure	+++	++	+	-	-
Venous drainage	++	++	+++	++	++
Atrial septum	++	+++	+	++	+
Membranes and baffles	+++	++	++	-	+
AV valves	+++	++	+	-	+
Fontan circulation	++	+++	++	+	++
Ventricular septum	+++	+	+	-	+
LV function	+++	+	++	++	+
RV function	++	+	+++	++	+
LV outflow tract	+++	++	+	-	++
RV outflow tract	+++	++	+	-	++
Pulmonary branches	+	-	+++	++	++
Pulmonary hypertension	++	-	-	+	+++
Qp:Qs	++	-	++	++	++
Sinus of Valsalva fistula	++	+++	++	++	+
Coarctation	++	+	+++	-	+
Aortic aneurysm	+	++	+++	-	+
Ductus or window	+++	++	+	++	+
Fistulas and collaterals	+	-	++	-	+++
Coronary anomalies	+	++	++	++	+++
Thrombosis	+	+++	++	-	-
Endocarditis	++	+++	-	-	-

+++ indicates diagnostic procedure of choice in most cases; ++, very useful procedure in most cases; +, not very useful procedure in most cases; – diagnostic procedure not indicated.

when TTE does not provide a complete anatomical or functional assessment. The information offered by these techniques may often be redundant, thus we should choose their sequence carefully to avoid unnecessary examinations. On other occasions, the information is complementary and therefore, we would have to resort to different diagnostic techniques to complete the evaluation. There are very few comparative studies of the clinical utility of these techniques.[43] Table 1 compares the diagnostic value of the different imaging techniques based on information in the medical literature and on daily clinical experience. The utility of each technique in the assessment of the different aspects of congenital heart disease has been classified into four groups: *a)* diagnostic procedure of choice in most cases; *b)* procedure of great utility in selected cases; *c)* procedure of limited utility in most cases, and *d)* procedure not indicated. This classification is similar to the indications of class I, IIa, IIb and III, though we have sought to avoid this nomenclature in order to separate personal opinion from well-established clinical guidelines. The main indications of the different third-level diagnostic techniques according to this classification are shown in Table 2.

REFERENCES

1. Moller JH, Taubert KA, Allen HD, Clark EB, Lauer RM, a Special Writing Group from the Task Force on Children and Youth, American Heart Association. Cardiovascular health and disease in children: current status. Circulation 1994;89:923-30.
2. Oliver J. Cardiopatías congénitas del adulto: residuos, secuelas y complicaciones de las cardiopatías congénitas operadas en la infancia. Rev Esp Cardiol 2003;56:73-88.
3. Perfof JK, Warnes CA. Challenges posed by adults with repaired congenital heart disease. Circulation 2001;103:2637-43.
4. Stumper OF, Sreeram N, Elzenga NJ, Sutherland GR. Diagnosis of atrial situs by transesophageal echocardiography. J Am Coll Cardiol 1990;16:442-6.
5. Applegate KE, Goske MJ, Pierce G, Murphy D. Situs revisited: imaging of the heterotaxy syndrome. Radiographics 1999;19:837-52.
6. Becker AE, Anderson RH. Pathology of congenital heart disease. 1. Basic approach and philosophy. Postgraduate Pathology Series 1983;3-27.
7. Seward JB. Biplane and multiplane transesophageal echocardiography: evaluation of congenital heart disease. Am J Card Imaging 1995;9:129-36.
8. White CS, Baffa JM, Haney PJ, Pace ME, Campbell AB. MR imaging of congenital anomalies of the thoracic veins. Radiographics 1997;17:595-608.
9. Ferrari VA, Scott CH, Holland GA, Axel L, Sutton MS. Ultrafast three-dimensional contrast-enhanced magnetic resonance angiography and imaging in the diagnosis of partial anomalous pulmonary venous drainage. J Am Coll Cardiol 2001;37:1120-8.
10. Hausmann D, Daniel WG, Mügge A, Ziemer G, Pearlman AS. Value of transesophageal color Doppler echocardiography for detection of different types of atrial septal defect in adults. J Am Soc Echocardiogr 1992;5:481-8.

TABLE 2. **Main indications for secondary imaging techniques**

Imaging technique	Main indications
Transesophageal echocardiography	Patients with very limited transthoracic window
	Atrial situs and segment ordering in complex cases
	Patent foramen ovale and atrial septal aneurysm
	ASD closure with percutaneous device
	Sinovenous ASD
	Cor triatriatum
	Dehiscence or stenosis of intraatrial baffles
	Complications of Fontan surgery
	Criteria for operability of Ebstein anomaly
	Perioperative assessment of mitral, aortic or tricuspid regurgitation
	Differential diagnosis of subaortic stenosis
	Fistulas of the sinus of Valsalva
	Ductus arteriosus with pulmonary hypertension
	Suspected infectious endocarditis
	Thromboembolic complications
Magnetic resonance imaging	Visceroatrial situs in heteroataxia syndromes
	Pulmonary or systemic venous drainage anomalies
	Size and function of right pulmonary or systemic ventricle
	Stenosis or hypoplasia of pulmonary trunk or branches
	Native or postoperative aortic coarctation
	Aneurysms of ascending or descending aorta
	Anomalies of aortic arch and arterial rings
	Surgical fistulas and systemic collaterals
Nuclear cardiology	Determination of Qp:Qs
	Ventricular size and function (left or right)
	Pulmonary perfusion in stenosis or hypoplasia of pulmonary branches
	Myocardial ischemia in coronary anomalies
Cardiac catheterization and angiocardiography	Pulmonary blood pressure, vascular resistance and response to vasodilators in patients with acute pulmonary hypertension
	Congenital coronary anomalies
	Arteriovenous or bronchial fistulas
	Systemic collaterals prior to unifocalization
	Percutaneous therapeutic procedures
	Preoperative procedure in complex heart disease
	Coronary arteriography in patients over 50 years or with risk factors for ischemic heart disease

11. Oliver JM, Gallego P, González A, Domínguez FJ, Aroca A, Mesa JM. Sinus venosus syndrome: atrial septal defect or anomalous venous connection? A multiplane transoesophageal approach. Heart 2002;88:634-8.
12. Rosenfeld HM, van der Velde ME, Sanders SP, Colan SD, Parness IA, Lock JE, et al. Echocardiographic predictors of candidacy for successful transcatheter atrial septal defect closure. Cathet Cardiovasc Diagn 1995;34:29-34.
13. Evangelista A, Aguadé S, Candell-Riera J, Ángel J, Galve E, García del Castillo H, et al. Cuantificación del cortocircuito de la comunicación interauricular mediante oximetría, isótopos y ecocardiografía Doppler. ¿Existe un método de referencia? Rev Esp Cardiol 1998;51(Suppl 1):2-9.
14. Mas JL, Arquizan C, Lamy C, Zuber M, Cabanes L, Derumeaux G, et al. Recurrent cerebrovascular events associated with patent foramen ovale, atrial septal aneurysm, or both. N Engl J Med 2001;345:1740-6.
15. Schluter M, Langenstein BA, Thier W, Schmiegel WH, Krebber HJ, Kalmar P, et al. Transesophageal two-dimensional echocardiography in the diagnosis of cor triatriatum in the adult. J Am Coll Cardiol 1983;2:1011-5.
16. Kaulitz R, Stumper OF, Geuskens R, Sreeram N, Elzenga NJ, Chan CK, et al. Comparative values of the precordial and transesophageal approaches in the echocardiographic evaluation of atrial baffle function after an atrial correction procedure. J Am Coll Cardiol 1990;16:686-94.
17. Sreeram N, Stümper OFW, Kaulitz R, Hess J, Roelandt JRTC, Sutherland GR. Comparative value of transthoracic and transesophageal echocardiography in the assessment of congenital abnormalities of the atrioventricular junction. J Am Coll Cardiol 1990;16:1205-14.
18. Hsu YH, Santulli T Jr, Wong AL, Drinkwater D, Laks H, Williams RG. Impact of intraoperative echocardiography on surgical management of congenital heart disease. Am J Cardiol 1991;67:1279-83.
19. Danielson GK, Driscoll DJ, Mair DD, Warnes CA, Oliver WC Jr. Operative treatment of Ebstein's anomaly. J Thorac Cardiovasc Surg 1992;104:1195-202.
20. Stumper O, Sutherland GR, Geuskens R, Roelandt JR, Bos E, Hess J. Transesophageal echocardiography in evaluation and management after a Fontan procedure. J Am Coll Cardiol 1991;17:1152-60.
21. Fyfe DA, Kline CH, Sade RM, Gillette PC. Transesophageal echocardiography detects thrombus formation not identified by transthoracic echocardiography after the Fontan operation. J Am Coll Cardiol 1991;18:1733-7.

22. Sampson C, Martínez J, Rees S, Somerville J, Underwood R, Longmore D. Evaluation of Fontan's operation by magnetic resonance imaging. Am J Cardiol 1990;65:819-21.
23. Fogel MA, Donofrio MT, Ramaciotti C, Hubbard AM, Weinberg PM. Magnetic resonance and echocardiographic imaging of pulmonary artery size throughout stages of Fontan reconstruction. Circulation 1994;90:2927-36.
24. Nichols K, Saouaf R, Ababneh AA, Barst RJ, Rosenbaum MS, Groch MW, et al. Validation of SPECT equilibrium radionuclide angiographic right ventricular parameters by cardiac magnetic resonance imaging. J Nucl Cardiol 2002;9:153-60.
25. Lorenz CH, Walker ES, Graham TP Jr, Powers TA. Right ventricular performance and mass by use of cine MRI late after atrial repair of transposition of the great arteries. Circulation 1995; 92(Suppl):II233-9.
26. Helbing WA, Niezen RA, Le Cessie S, van der Geest RJ, Ottenkamp J, de Roos A. Right ventricular diastolic function in children with pulmonary regurgitation after repair of tetralogy of Fallot: volumetric evaluation by magnetic resonance velocity mapping. J Am Coll Cardiol 1996;28:1827-35.
27. Oliver JM, González A, Gallego P, Sánchez-Recalde A, Benito F, Mesa JM. Discrete subaortic stenosis in adults: increased prevalence and slow rate of progression of the obstruction and aortic regurgitation. J Am Coll Cardiol 2001;38:835-42.
28. Boxer RA, Fishman MC, LaCorte MA, Singh S, Parnell VA Jr. Diagnosis and postoperative evaluation of supravalvular aortic stenosis by magnetic resonance imaging. Am J Cardiol 1986;58:367-8.
29. Gallego P, Castro A, Oliver JM, Barquero JM, Díaz Infante E, Araji O, et al. Anatomía funcional de la regurgitación aórtica. Implicaciones del diagnóstico con ecocardiografía transesofágica en la reparación valvular. Resultados iniciales. Rev Esp Cardiol 2002;554(Supl 2):53.
30. Kucher N, Seiler C, Allemann Y, Eberli FR. Double-chambered right ventricle. Circulation 2001;103:105-6.
31. Oliver JM, Garrido A, González A, Benito F, Mateos M, Aroca A, et al. Rapid progression of mid-ventricular obstruction in adults with double-chambered right ventricle [en prensa]. J Thorac Cardiovasc Surg, 2003.
32. Geva T, Greil GF, Marshall AC, Landzberg M, Powell AJ. Gadolinium-enhanced 3-dimensional magnetic resonance angiography of pulmonary blood supply in patients with complex pulmonary stenosis or atresia: comparison with x-ray angiography. Circulation 2002;106:473-8.
33. Fogel MA, Hubbard A, Weinberg PM. A simplified approach for assessment of intracardiac baffles and extracardiac conduits in congenital heart surgery with two- and three-dimensional magnetic resonance imaging. Am Heart J 2001;142:1028-36.
34. Oshinski JN, Parks WJ, Markou CP, Bergman HL, Larson BE, Ku DN, et al. Improved measurement of pressure gradients in aortic coarctation by magnetic resonance imaging. J Am Coll Cardiol 1996;28:1818-26.
35. Therrien J, Thorne SA, Wright A, Kilner PJ, Somerville J. Repaired coarctation: a «cost-effective» approach to identify complications in adults. J Am Coll Cardiol 2000;35:997-1002.
36. Berdon WE. Rings, slings, and other things: vascular compression of the infant trachea updated from the midcentury to the millennium-the legacy of Robert E. Gross, MD, and Edward B. D. Neuhauser, MD. Radiology 2000;216:624-32.
37. Galeote G, Oliver JM, Domínguez FJ, Fuertes J, Calvo L, Sobrino JA. Seudocoartación aórtica complicada con seudoaneurisma gigante. Rev Esp Cardiol 2000;53:287-9.
38. Garrido Martín A, Oliver Ruiz JM, González AE, Mesa García JM, Benito F, Sobrino Daza JA. Ecocardiografía transesofágica multiplana en la valoración preoperatoria de fístulas de senos de Valsalva en cavidades derechas. Rev Esp Cardiol 2002;55:29-36.
39. Shyu KG, Lai LP, Lin SC, Chang H, Chen JJ. Diagnostic accuracy of transesophageal echocardiography for detecting patent ductus arteriosus in adolescents and adults. Chest 1995;108:1201-5.
40. Duerinckx A, Atkinson D, Klitzner TS, Perloff J, Drinkwater D, Laks H. MR imaging of surgical complications of systemic-to-pulmonary artery shunts. Magn Reson Imaging 1996;14:1099-105.
41. Feinstein JA, Moore P, Rosenthal DN, Puchalski M, Brook MM. Comparison of contrast echocardiography versus cardiac catheterization for detection of pulmonary arteriovenous malformations. Am J Cardiol 2002;89:281-5.
42. Vliegen HW, Doornbos J, de Roos A, Jukema JW, Bekedam MA, van der Wall EE. Value of fast gradient echo magnetic resonance angiography as an adjunct to coronary arteriography in detecting and confirming the course of clinically significant coronary artery anomalies. Am J Cardiol 1997;79:773-6.
43. Hirsch R, Kilner PJ, Connelly MS, Redington AN, St John Sutton MG, Somerville J. Diagnosis in adolescents and adults with congenital heart disease. Prospective assessment of individual and combined roles of magnetic resonance imaging and transesophageal echocardiography. Circulation 1994;90:2937-51.