UPDATE

Congenital heart disease in adults: The contribution of multidetector CT

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Abstract

Congenital heart disease is relatively common among adults. Patients’ conditions have generally been diagnosed previously and imaging tests are requested for follow-up or for complications of the anomaly or of its surgical correction. Classically, these patients were studied with echocardiography and cardiac catheterization, but multidetector CT and magnetic resonance imaging have changed the approach because these techniques show the anatomy of heart defects and their correction very clearly.

We emphasize the importance of multidetector CT as a complementary technique for the study of congenital heart disease that is newly discovered in adults or for the follow-up of congenital heart disease that was surgically corrected during childhood. When vascular anomalies are present outside the heart or after palliative surgery, multidetector CT shows anatomical details that are difficult or impossible to see with echocardiography. We also emphasize the frequent association between pulmonary hypertension and congenital heart disease that can debut in adults.

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KEYWORDS

Heart defects congenital;
Heart septal defects atrial;
Heart septal defects ventricular;
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Tetralogy of Fallot;
Transposition of great vessels;
Spiral cone-beam CT;
Computed tomography multidetector

PALABRAS CLAVE

Cardiopatías congénitas;
Comunicación interauricular;
Comunicación interventricular;

Resumen

Las cardiopatías congénitas (CC) constituyen actualmente una enfermedad no infrecuente en el adulto. Generalmente el paciente está diagnosticado previamente y el estudio se solicita como seguimiento o por complicaciones de la anomalía o de su corrección quirúrgica.

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Congenital heart disease in adults: The contribution of multidetector CT

Introduction

Congenital heart disease (CHD) is a relatively common disease. Thanks to the success of surgery in the past 30 years, which has resulted in greater survival and better quality of life, more and more cases are now seen in adults. According to the European Congenital Heart Disease Organization (ECHDO), before surgery was available, less than 20% of children born with CHD reached adulthood. Now, the majority of deaths by CHD are observed in adults. Due to the fact that many patients with CHD are monitored into the 4th or 5th decade of their lives, and because there is simple CHD that manifests in the adulthood, it is predicted that there will soon be more adults than children with CHD.1,2

Current imaging techniques, especially multidetector computerized tomography (MDCT) and magnetic resonance imaging (MRI), present advantages over traditional methods of radiographic study, such as cardiac angiography and echocardiography, which results in a greater demand for those tests. MDCT and MRI permit a complete study of the cardiac anatomy, mediastinal and pulmonary vessels and thoracic or abdominal structures that can form part of the congenital changes, with short scanning times and small contrast media doses. Moreover, they allow for an assessment of surgical result and complications.

To offer useful information to the cardiologist and cardiac surgeon, the radiologist ought to know the basic anatomy and physiology of CHD before and after surgical repair, as well as understand the history and clinical signs of the patient that motivated the radiology study. When MDCT is used as a method of study, the radiologist ought to be aware that the study is purely anatomical and complementary to ultrasound, which offers a functional study.3

This work does not intend to be an exhaustive study of CHD, but rather attempts to present the most representative cases that we have been able to study and emphasizes the important contribution of MDCT in the multidisciplinary study of the adult patient with CHD.

Study techniques

Echocardiography

Transthoracic echocardiogram is the preferred imaging technique for the study of CHD because it combines the analysis of cardiac structure and function through two-dimensional imaging with a Doppler hemodynamic assessment (continuous, pulsed, color and tissue).

Moreover, in selected cases in which there are diagnostic doubts or the anatomy should be well described for intervention procedures, a transesophageal echocardiogram is of vital importance because imaging quality drastically improves.

However, despite advances in ultrasound, including the emergence of the second harmonic, ultrasound contrasts or three-dimensional ultrasound which is still in its developmental stages, patients with complex CHD have various limitations including suboptimal imaging quality due to a poor ultrasonic window due to previous cardiac surgeries or the need to assess extracardiac structures such as systemic pulmonary fistulas or cavopulmonary connections. Therefore, other imaging techniques like cardiac MRI or MDCT are necessary to complement known information and to make more accurate diagnostic and therapeutic decisions.

Magnetic resonance imaging

MRI has good spatial and temporal resolution and allows for a functional volumetric and flow assessment as well as tissue characterization. The main drawbacks are the longer scanning times, which in children require sedation or even general anesthesia; and also the imaging artifacts that cardiac prostheses for CHD correction can cause.

Cardiac catheterization

Currently, cardiac catheterization is rarely used as a diagnostic tool and is generally performed when hemodynamic assessment or percutaneous interventions are required.

Multidetector computerized tomography

MDCT is a fast imaging technique that has excellent spatial resolution, higher than MRI, thanks to a thinner collimation leading to isotropic voxels. However, temporal resolution is lower than MRI and, accordingly, it is not usually applied for ventricular function analysis. MDCTs most important drawback, the radiation dose, can be reduced by decreasing the kilovoltage according to the size and age of the patient. In studies with retrospective gating, the milliamperage of
the tube current can be modulated with different commercially available procedures, and new techniques of prospective cardiac synchronization also permit a significant reduction of the radiation dose.\(^3\,^4\)

MDCT offers the advantage of a complete anatomical study of complex congenital anomalies, not only of the heart itself, but also of the mediastinal vessels, tracheobronchial tree, lung parenchyma, chest wall, diaphragm and upper abdomen. In addition, MDCT allows for an adequate assessment of intracardiac or vascular devices and for characterization of the functional status or obstruction of the anastomoses.

**Scanning technique with multidetector computerized tomography**

In the cases we present in this review, MDCT, both with 16 and 64 channels, used the following parameters: collimation of 16 × 0.75 mm or 64 × 0.62 mm, for reconstruction of 1 mm slices with intervals of 0.5 mm (50% overlap) and a gantry rotation time of 0.5 sec.

Data acquisition was achieved without cardiac gating synchronization in the craniocaudal direction, with medium or light breath holding, 120 kV, 200-250 mA and automatic dose regulation.

A bolus of 70 to 90 cm\(^3\) non-ionic iodinated contrast media (350 mg/ml iodine) was injected in the antecubital vein at a rate of 4 cm\(^3\)/sec, followed by a 40 cm\(^3\) bolus of intravenous (IV) saline at the same injection rate. As a result, a uniform contrast bolus is achieved with a smaller total amount of contrast media. The bolus tracking technique was used to optimize the initiation of the data acquisition; the region of interest (ROI) was placed in the anatomical structure that we wanted to study in greater detail and we set a threshold of 180 Hounsfield units to study cavities or structures on the right heart and of 150 Hounsfield units for structures on the left heart.

The post-processing was performed in a dedicated working station with multiplanar reformatting, maximum intensity projections (MIP) and volumetric reconstructions, with different windows settings for the lung parenchyma (W1600 L-600), mediastinum (W500 L35) and vasculature (W700 L100).

The scanning time is very brief, between 8-12 s, and the reconstructions are not difficult if the anatomy or the surgical procedure performed on the patient is known. In addition, scanning with MDCT is very safe in the presence of metallic coils, pacemakers, valvular or tubular prostheses, or vascular clips. Another significant advantage of MDCT is the broad implementation of this type of equipment in hospitals and clinics, so the technical aspects and diagnostic methods are already known by the majority of radiologists.

**Atrial septal defects**

Overall, atrial septal defects (ASD) are the most frequent congenital heart defects in adults (30%).\(^5\,^6\,^7\) To understand...
Congenital heart disease in adults: The contribution of multidetector CT

Development of the interatrial septum

In the early stages of embryonic development, the right and left atria form a common cavity. Once the endocardial cushions are developed inside the atrioventricular canal, the division of the atria begins with the formation of the septum primum, a crest of tissue that grows from the posterior and superior wall of the atrial cavity and moves toward the endocardial cushions. The space that remains between the free margin of the septum primum and the endocardial cushions is called the ostium primum (fig. 1A). Before complete closure of the ostium primum, the uppermost portion of the septum primum is pierced, forming the ostium secundum. Subsequently, on the right side of the septum primum, a tissue develops that covers the ostium secundum and forms the foramen ovale. This tissue, called the septum secundum, is formed by a fold of the right atrial wall between the superior vena cava and the right pulmonary veins (fig. 1B).

In the development of the fetus, there is a shunting of blood from the right to the left atrium through the foramen ovale due to increased pressure in the right atrium caused by the elevated vascular resistances in the pulmonary arterial vessels. At birth, the increase of pulmonary blood flow elevates the pressure in the left atrium and causes the closure of the foramen ovale; what remains in this area of the interatrial septum is a thin oval area called the fossa ovalis.

Types of atrial septal defects:

1. **Ostium secundum defect**: It is the most frequent ASD (75%) and is located in the fossa ovalis, in the middle portion of the septum. It is the result of an incomplete growth of the septum secundum or an excessive resorption of the septum primum. It is normally a single defect although fenestrated defects are also possible (fig. 2).

2. **Ostium primum defect**: This defect appears in 15% of cases and is located in the lower portion of the interatrial septum. It is produced by a defective fusion of the septum primum and the endocardial cushions. It is considered the most simple form of atrioventricular septal or atrioventricular canal defect (fig. 3).

3. **Sinus venosus defect**: This atrial defect occurs in 10% of cases and it is the result of an abnormal fusion between the embryonic sinus venosus and the atrial cavity. It mainly affects the insertion of the inferior vena cava (fig. 4).

4. **Coronary sinus defect**: It is the less frequent of the ASD (less than 1% of cases) resulting from a direct communication of the coronary sinus with the left atrium. The existence of a dilated coronary sinus frequently indicates this type of septal defect.

Figure 3  Forty one-year-old male diagnosed with a partial defect of the atrioventricular canal. On the echocardiography, an increase in size and wall thickening of the right ventricle was visualized, as well as an interventricular septal defect near the junction of the tricuspid and mitral valves. A) The image shows the ostium primum type of atrial septal defect (red arrow) and an aneurysm in the membranous portion of the interventricular septum (yellow arrow). B) The CT scan was performed for preoperative evaluation. The axial oblique slice identifies the same septal defects than in the ultrasound examination (green arrows mark the atrioventricular valves).

Figure 4  Fifteen-year-old female being studied for severe pulmonary hypertension. A) The transthoracic echocardiography showed an increase in size of the atrium, right ventricles and pulmonary artery, with severe right ventricular dysfunction and an increase of pulmonary pressures. After the contrast injection (agitated saline solution), a left-to-right shunt was observed in the upper portion of the atrial septum. The transesophageal echocardiogram confirmed a sinus venous atrial septal defect with normal drainage of the left pulmonary veins. Given the difficulty of assessing the drainage of the right pulmonary veins, a CT scan was performed. B) Oblique coronal image that shows a posterior and superior atrial septal defect (arrow) close to the insertion of the superior vena cava. C) Axial MIP reconstruction identifies the anomalous connection between the right upper pulmonary vein (orange arrow) with the posterior wall of the superior vena cava draining in the right atrium, and the atrial septal defect (green arrow).
Associated injuries

Atrial septal defects can be associated with other cardiac malformations in nearly 30% of all cases. The partial anomalous pulmonary venous drainage is associated with ASD in 80-90% of cases, primarily with defects of the sinus venosus (fig. 4C) and less frequently with ostium secundum and coronary sinus defects. Defects of the coronary sinus can be associated with total anomalous pulmonary drainage and with a persistent left superior vena cava.

Other frequent associations are valvular heart disease, like stenosis of the pulmonary or mitral valve, and prolapse of the mitral valve (fig. 2B). Ventricular septal defects, patent ductus arteriosus and coarctation of the aorta may also be associated.

Evolution and treatment of atrial septal defects

In atrial septal defects, there is often a left-to-right shunt due to the normal higher pressure in the left cavities, as a result the lungs and right cardiac cavities suffer a volume overload that lead to signs and symptoms of right heart failure (recurrent lung infections, fatigue and intolerance to exercise, supraventricular arrhythmias, paradoxical embolism, stroke, etc.). A common long-term consequence of this shunting is the development of pulmonary arterial hypertension. Therefore, it is important to look for pulmonary arterial hypertension signs when evaluating patients with ASD, in addition to assessing the integrity of the interatrial septum in patients being studied for pulmonary arterial hypertension. If signs of pulmonary arterial hypertension are identified on CT scan, it is necessary to perform an echocardiography to confirm its existence.

Pulmonary arterial hypertension can occasionally (5%) produce an inversion of the shunt direction (right-to-left) known as Eisenmenger’s syndrome. In these cases, the ASD repair is not indicated.

Likewise, in patients with ASD and pulmonary arterial hypertension, a careful assessment of the venous pulmonary drainage is required due to the frequent association between these anomalies (fig. 5).

Ultrasound assessment of ASD is essential for determining the surgical, percutaneous or medical treatment of patients, and it is important to define the size and location of the

Figure 5 Twenty-nine-year-old female with a previously corrected atrial septal defect of the upper sinus venosus. A CT scan was performed due to suspicion of an anomalous venous drainage. Coronal volumetric reconstruction shows the drainage of the right upper and middle pulmonary veins in the posterior wall of the superior vena cava. In patients with atrial septal defects and arterial pulmonary hypertension, an adequately assessment of the pulmonary venous drainage is warranted as it is frequently associated with those anomalies.

Figure 6 Forty-year-old male with a percutaneous closure of an atrial septal defect by an Amplatzer device. Transthoracic echocardiography six months after the procedure revealed persistence of the interatrial shunt. A partial detachment of the Amplatzer was suspected and a MDCT scan was requested.

A) The Axial oblique image shows the persistent atrial septal defect (red arrow) and the Amplatzer device protruding into the right atrium (green arrow). B) The volumetric reconstruction shows the oblique position of the Amplatzer with respect to the atrial septum (parallel to the yellow dashed line).

Figure 7 Schematic drawing of the two components of the interventricular septum: membranous and muscular septa.
Congenital heart disease in adults: The contribution of multidetector CT

septal defect, the associated anomalies and any signs of pulmonary arterial hypertension. CT is a useful tool for the study of associated lesions and complications (fig. 6).

**Ventricular septal defects**

Defects of the interventricular septum constitute the most frequent CHDs at birth (20%). However, the incidence in adults is much lower due to the fact that many small defects close spontaneously.

**Types of ventricular septal defects:**

There are four types of ventricular septal defects (VSD), classified according to the affected portion of the septum. The most frequent are perimembranous defects (75%), which are situated in the membranous portion of the septum, at the level of the outflow tract of the left ventricle below the aortic valve. Due to their location, they are also called subaortic defects (figs. 8A and 9A).

**Associated lesions**

Although VSD are frequently isolated lesions, they represent a common component of complex anomalies such as conotruncal defects (Tetralogy of Fallot and transposition of the great vessels [TGV]), a double outlet right ventricle and complete atrioventricular defects. The principal reason for requesting an MDCT is to evaluate the presence of associated complications.

Other possible associations, especially with conal or supracristal defects, include the following:

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**Figure 8** Thirty six-year-old female with hemoptysis and history of ventricular septal defect. The ultrasound examination (not shown) showed a severe pulmonary arterial hypertension, with a large ventricular septal defect, predominantly right-to-left shunt (Eisenmenger’s syndrome) and a dilated dysfunctional right ventricle. A CT examination was requested to assess both the ventricular septal defect and the hemoptysis. A) The axial oblique slice shows the ventricular septal defect in the upper portion of the interventricular septum. B) Volumetric reconstruction demonstrates a coarctation of the aorta (arrow).

**Figure 9** Twenty two-year-old female with history of subaortic ventricular septal defect and valvular and infundibular pulmonary atresia. Systemic pulmonary collateral vessels supplied the pulmonary flow. The ultrasound examination (not shown) revealed an absent pulmonary valve and hypoplasia of the trunk of the pulmonary artery. A CT scan was carried out to evaluate the pulmonary vascularization. A) The oblique coronal slice confirms the ventricular septal defect (arrows) with an enlarged ascending aorta. B) Axial MIP reconstruction shows the absence of the trunk and main branches of the pulmonary artery. C) Coronal MIP reconstruction. The bronchial arteries sprout from the descending aorta (arrows) and connect with the pulmonary lobar arteries.

**Figure 10** The schematic drawing shows A) an Ebstein’s anomaly and, B) a normal right ventricle.
- Severe forms of obstruction to aortic outflow (coarctation of the aorta, interrupted aortic arch, subaortic stenosis), frequent in large defects (fig. 8B).
- Progressive prolapse of the aortic valvular tissue that gives way to a progressive distortion of the aortic valve or sinus of valsalva, which can lead to aortic valve insufficiency, development of an aneurysm of the sinus of valsalva or formation of a fistula between the right ventricle and the right coronary sinus of Valsalva.
- Infundibular or valvular pulmonary stenosis.
- Valvular or subvalvular aortic stenosis.
- Patent ductus arteriosus.
- Pulmonary atresia (figs. 9B,C).

\section*{Ebstein’s anomaly}

Ebstein’s anomaly is an infrequent CHD with an extremely variable clinical presentation and represents approximately 0.5-1\% of the total cases of CHD.\textsuperscript{21} It is characterized by varying grades of dysplasia and downward displacement of the septal and posterior leaflets of the tricuspid valve into the inflow portion of the right ventricle.\textsuperscript{22} The anterior valve, although in normal position, is usually long and redundant. Moreover, the tricuspid annulus is dilated and displaced inferiorly, which leads to a significant increase in the size of the right atrium at the expense of a reduction in the size of the right ventricle (atrialization of the ventricle). Accordingly, the right ventricle mechanical function is impaired and facilitates the development of heart failure (figs. 10 and 11).

In adults, symptoms are typically a consequence of tricuspid regurgitation and ventricular dysfunction. Arrhythmias represent a significant cause of morbidity and sudden death at any age.\textsuperscript{23}

Even if the diagnosis of Ebstein’s anomaly is based on ultrasound findings, anatomical studies using this technique are limited in some cases, and MDCT and MRI provide essential diagnostic clues for therapeutic treatment.\textsuperscript{24} Carpentier’s classification, based on the valvular displacement grade and on the valvular motility of the right ventricle, is used to quantify the disease and is very useful for deciding the surgical technique.\textsuperscript{25}

The different surgical techniques available are intended for the plication of the atrialized right ventricle, the correction or substitution of the valvular anomaly or the performance of palliative techniques related to pulmonary circulation (fig. 12C).\textsuperscript{26}

![Figure 11](image1.png)

\textbf{Figure 11} Thirty eight-year-old male diagnosed with an Ebstein’s anomaly, atrial septal defect of the ostium secundum type and pulmonary artery hypoplasia. Surgical repair was performed with closure of the atrial septal defect, a tricuspid valvuloplasty and a Glenn technique (anastomosis of the superior vena cava with the right pulmonary artery). Both A) MDCT and B) transthoracic ultrasound images show the tricuspid valve displacement into a shortened right ventricle. C) Axial and D) sagittal oblique MIP reconstructions showing the anastomosis between the superior vena cava and the right pulmonary artery. The patient presented with supraventricular tachycardia that was treated with radiofrequency ablation.
Associated lesions:
- Atrial septal defect (fig. 12A).27
- Pulmonary stenosis or atresia (fig. 12B).
- Tricuspid stenosis.
- Ventricular septal defect.
- Patent ductus arteriosus.
- Mitral stenosis or mitral valve prolapse.
- Bicuspid or atretic aortic valve.
- Tetralogy of Fallot.
- Corrected or partial TGV.28

Tetralogy of Fallot

Tetralogy of Fallot is one of the most frequent cyanotic CHDs in an adult (around 10% of the total) that is defined by four pathological findings (figs. 13 and 14A,B):

- Outflow obstruction of the right ventricle due to pulmonary stenosis, frequently infundibular.
- Hypertrophy of the right ventricle.
- Ventricular septal defect.
- Overriding aorta.

These alterations are a consequence of a single developmental change: the misalignment of the supraventricular crest associated with an underdevelopment of the infundibulum.

Associated lesions:
- Atrial septal defect (Pentalogy of Fallot).
- Right aortic arch.
- Atresia of the pulmonary artery (fig. 14C).
- Anomalous coronary artery (frequently consisting of a coronary artery of the cone that crosses the infundibulum and can hinder surgical correction) (fig. 14D).
- Agenesis or hypoplasia of the pulmonary valve with pulmonary insufficiency.
- Patent ductus arteriosus.
- Aortic insufficiency.
- Anomalous pulmonary venous drainage.
- Persistent left superior vena cava.
- Right subclavian artery.
- Collateral vessels in the aortopulmonary window.
- Rarely, tracheoesophageal fistula, rib anomalies and scoliosis.29

MDCT is especially useful in the assessment of complex heart diseases, such as the Tetralogy of Fallot, that may be associated with coronary and peripheral pulmonary artery anomalies, which in turn can determine the appropriate surgical technique. Moreover, MDCT has an essential role in the assessment of the postsurgical results.30

Generally, surgical technique depends on the age of the patient, the grade of cyanosis, the degree of obstruction to right ventricle outflow and of the pulmonary valvular stenosis, the size of the ventricular septal defect, the presence and size of the pulmonary arteries and any other associated anomalies.29 The treatment of choice is total early correction between 3-7 years of age. Another
alternative is the performance of a palliative technique like that of Blalock-Taussig in children that are not candidates for complete repair until the pulmonary arterial branches reach a size that permits surgical repair. The modified Blalock-Taussig technique permits the flow of blood from a subclavian artery to the ipsilateral pulmonary artery via a synthetic tube. In patients that cannot have the outflow tract of the right ventricle enlarged (for example, due to the existence of an anomalous coronary artery in that area), a duct can be created that goes from the right ventricle to the cone of the pulmonary artery to avoid obstruction.

Even though the prognosis of patients that have been treated surgically is favorable, residual effects, complications and sequelae are frequent.\textsuperscript{31-33}

**Transposition of the great vessels**

TGV is one of the most frequent cyanotic CHDs. The complete transposition of great arteries is characterized by a ventriculoarterial discordance such that the aorta originates in the morphologic right ventricle and situates itself anteriorly and to the right of the pulmonary artery, and the pulmonary artery originates in the morphologic left ventricle (D-TGV) (fig. 15). The systemic and pulmonary venous blood are mixed by a bidirectional shunt through a patent foramen ovale, patent ductus arteriosus or ventricular septal defect.

In some cases, an atrioventricular discordance develops in addition to the ventriculoarterial discordance, and, the left atrium is connected to the right ventricle (from which the aorta originates), while the right atrium is connected to the left ventricle (from which the pulmonary artery originates). Thus, the transposition is functionally corrected by the ventricular inversion and is called a congenitally corrected TGV. This represents less than 1% of the total cases of CHD.\textsuperscript{34}

Approximately 99% of patients with a corrected transposition have other associated anomalies\textsuperscript{35,36} such as the following:
Congenital heart disease in adults: The contribution of multidetector CT

- Ventricular septal defect (fig. 16).
- Outflow obstruction of the left ventricle.
- Coarctation of the aorta.\(^{37}\)
- Pulmonary stenosis (subvalvular, valvular, or supravalvular).\(^{38}\)
- Tricuspid valve anomalies, such as a dysplastic valve or Ebstein’s anomaly.
- Atrial septal defect.
- Patent ductus arteriosus
- Hypertrophy and dysfunction of the right ventricle.

Figure 15  Schematic drawing showing the ventriculoarterial discordance in the transposition of the great vessels.

Figure 16  Transposition of the great vessels. MDCT. The sagittal-oblique MIP reconstruction shows an anterior aorta, arising from the right ventricle, and a posterior pulmonary artery, which originates in the left ventricle. In the transposition of the great vessels the aorta and the pulmonary artery exhibit a characteristic parallel course. Ventricular septal defect (green arrows) and stenosis of the pulmonary artery trunk secondary to a banding procedure (red arrow).

Figure 17  Twenty seven-year-old male with transposition of the great vessels, tricuspid atresia, hypertrophied right ventricle, infundibular subpulmonary stenosis and a small ventricular septal defect. The patient presented with an atrial septal defect secondary both to a Rashkind procedure secondary to a Rashkind procedure, and also presented a Blalock-Taussig shunting. A MDCT was performed to evaluate the shunt function. The A) coronal and, B) volumetric reconstructions show the atrial septal defect (red arrow) and shunt permeability between the subclavian artery and the principal left pulmonary artery (yellow arrow).

- Anomalies in the anatomy of the coronary arteries that could determine the surgical technique.\(^{39}\) Thus, a detailed pre-surgical study of the coronary anatomy is paramount if an atrial or arterial switch is considered.\(^{40}\)
- Dextrocardia.

Regarding TGV treatment, there are two main procedures: the atrial switch (Mustard or Senning) that has been employed for years and the arterial switch, which is currently used. In the congenitally corrected transposition, patients that do not have other anomalies or only have minor associated anomalies generally do not require surgical repair. In cyanotic patients or in those that develop heart insufficiency or severe pulmonary stenosis, a double switch is performed in which the atroventricular and ventriculoarterial connections are exchanged. Associated anomalies, like a ventricular septal defect and pulmonary stenosis, generally are corrected in the same procedure\(^{37}\) (figs. 16 and 17).

Heterotaxy syndrome

Heterotaxy syndrome includes a wide spectrum of anomalies consisting of a developmental alteration of cardiac asymmetries or an abnormal arrangement of thoracoabdominal organs across the left-right axis of the body (fig. 18). The abnormalities related to the internal disposition of organs can be divided into the following subcategories:

- Situs solitus: normal disposition of organs and vessels in the body. The rate of CHD is only 0.6-0.8 %.\(^{41}\)
- Situs inversus: mirror-imaged arrangement of the internal organs along the left-right axis. The right or systemic atrium, which receives blood from the inferior vena cava,\(^{42}\) is situated in the left side of the body and is
accompanied by a three-lobed left lung, as well as the liver, the gallbladder and the inferior vena cava located on the left side. The morphological left atrium is situated on the right side, with a bilobed right lung, stomach, spleen and aorta all on the right side. The rate of CHD is 3-5%. Kartagener’s syndrome, which is associated with situs inversus, nasal polyposis and bronchiectasis, is present in 20% of patients with situs inversus.

- Situs ambiguous: includes any other alteration of the left-right axis. If the liver is symmetric and central, there...
is the possibility of bilateral symmetry with two right sides (asplenia syndrome, right isomerism or Ivemark syndrome), or with two left sides (polysplenia syndrome or left isomerism). The rate of CHD in these patients is very high, between 50-100%. CHDs associated with heterotaxy syndrome includes a broad spectrum of anomalies: 47

- Atrioventricular discordance.
- TGV.
- Anomalies in the systemic and pulmonary venous drainage.
- Atrial/ventricular septal defects (the most frequent anomalies, between 60 and 100% of the total) (fig. 19).
- Subpulmonary or aortic obstruction.

Conclusions

MDCT has become a useful imaging technique in the pre-surgical and postsurgical assessment of a great variety of CHDs in adults. This technique is complementary to echocardiography, MRI and cardiac catheterization due to its high spatial resolution, the ability to perform multiphase and functional studies, the speed and safety in the presence of pacemakers, valvular prostheses and other surgical devices. However, its main contributions in the study of CHDs in adults are the assessment of complex cardiopathies, vascular anomalies, extracardiac shunts and the assessment of postsurgical results.

Conflict of interest

The authors declare no conflict of interest.

Authorship

María Navallas has contributed to the conception and design of the study, to the analysis and to the interpretation of data; she has been involved in drafting the work and its critical revisions and has made relevant intellectual contributions.

Paula Orenes has contributed to the conception and design of the study and to the analysis and interpretation of the data; she has been involved in the drafting of the work and in its critical revision and has made relevant intellectual contributions.

María Antonia Sánchez Nistal has contributed to the conception and design of the study, to the collection of the data and to its analysis and interpretation. She has contributed in drafting the work and in its critical revision and has made relevant intellectual contributions. She has given her final approval to the version sent to be published.

Carmen Jiménez López Guarch has contributed to drafting the work and has made relevant intellectual contributions.

All the authors have read and approved the final version of the article.

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