Pictorial essay

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Echocardiography and imaging investigation in congenital cardiovascular anomalies – competition or complementarity? Part I: non-cyanogenic cardiovascular malformations

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Abstract

Echocardiography is the first technique used for diagnosing cardiovascular malformations (CVM). The results are often completed with multi-detector computer tomography (MDCT) and/or magnetic resonance imaging (MRI) for confirming/invalidating an abnormal pulmonary venous return in the case of atrial septal defect (ASD) or for the exact interpretation of cardiac function in pre/post-surgery cases with ASD, ventricular septal defect (VSD), and Fallot tetralogy. MDCT and MRI play an important role in the precise and complete diagnosis of Fallot tetralogy, in the anomalies of the right heart cavities and the arterial and venous pulmonary tree, the anomalies of the emergence and course of the coronary arteries, aortic coarctation and developmental anomalies of the aortic arch and supraaortic trunks. The complementarity of echocardiography with MDCT and MRI, in order to obtain details and to avoid invasive procedures and also the cooperation between the pediatrician, cardiologist, surgeon and radiologist, represent the key to the diagnosis and treatment of cardiovascular malformations, for the benefit of the patient.

Keyword: cardiovascular malformation, echocardiography, multidetector computer tomography, magnetic resonance imaging.

Introduction

Congenital cardiovascular malformations (CVM) are structural and functional abnormalities existing at birth, manifesting in utero, at birth, later, or never.

The CVM are a diagnostic and therapeutic challenge. Although their frequency is quite low, approximately 7-8 at 1000 births [1], precocious diagnosis and treatment are mandatory. The progress of cardiovascular surgery has changed the prognosis of these diseases, increasing the percentage of survival and the life quality of these patients.

For many years, the diagnosis of CVM implied standard radiography, catheterisation and angiography. The cardiac non-invasive imaging made spectacular progresses with the advent of echocardiography (ECHO), which substantially reduced cardiac catheterization.

The diagnosis of CVM is based on transthoracic echocardiography (TTE), sometimes associated with transesophageal echocardiography (TEE). The quality and ease of performing these techniques are important for the pediatric population [2].

The purpose of this pictorial essay is to present and illustrate the relative diagnostic role of ECHO and of the imaging techniques in CVM.

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Examination techniques

In CVM, ECHO uses different techniques: M-mode, grey-scale, color, continuous and pulsed wave Doppler, three-dimensional (3D) (in CVM complexes), transesophageal, intraoperative, with contrast products, and fetal. The transducers are adapted to the age and size of the patient. Sedation is, sometimes, necessary in toddlers.

ECHO of CVM requires a methodical, segmental and organized approach. Every structure is three-dimensional and must be explored on more views.

The contribution of MDCT in CVM is mainly morphological, although it can provide accurate functional information, as well. The spatial resolution achieved with high end devices allows a very precise study of even small cardiac and vascular structures. The speed of acquisition enables the examination of large volumes, within a short period of apnea. This limits artifacts and is very useful in the pediatric population [3-5]. Post-processing provides three-dimensional images able to deliver specific information. The main problem related to MDCT is patient irradiation.

MRI examination in CVM plays a key role [2-7]. MRI examination of the heart and great vessels implies high-end devices: magnetic field strength of 1.5-3T, ECG gating, diaphragm movement detectors, specific coils, MRI dedicated electrodes, and automated injectors. The commonly used sequences for heart exploration are: "cine" – SSFP, "black blood" – DIRFSE, "volume flow" – cine-phase contrast, angio MRI and perfusion IR EGT1. There is no standardized protocol, due to the diversity of possible morphological abnormalities in CVM. The currently available sequences provide dynamic images, synchronized with cardiac contractions and high-speed flow sequences with parallel imaging techniques that enable proper assessment of heart function. Gadolinium enhanced angio MRI sequences with peripheral venous injection are crucial for the study of CVM.

The scan planes used in MRI and MDCT for cardiovascular examination are well defined and are related to those used in echocardiography, for a better design of the cardio-surgical therapeutic approach.

Cardiovascular malformations can be categorized by several criteria. According to the clinical criteria – occurrence or not of cyanosis – correlated with pathophysiological mechanisms, CVM are classified as:

A. non-cyanogenic:

a. with left-right shunt: atrial septal defect (ASD), ventricular septal defect (VSD), common atrioventricular canal, common arterial trunk, the persistence of the arterial channel, etc and

b. without shunt: aortic stenosis, pulmonary stenosis, aortic coarctation, mitral valve malformations.

B. cyanogenic: with pulmonary outflow obstruction: tetralogy of Fallot, pulmonary atresia, tricuspid atresia and pulmonary stenosis, and with pulmonary hypertension and Eisenmenger syndrome

C. mixed.

Shunts are abnormal communications between the left and right circulation, due to cardiac septal defects (ASD, VSD) or at the level of the great vessels (persistence of the arterial channel) or arteriovenous communications (pulmonary, systemic and myocardial - fistulas).

Shunts may be present at one or more levels, being necessary to consider the presence, location, direction and size. The presence of a shunt is initially suggested by the dilatation of the cavity receiving the shunt. Shunts can be viewed directly, as a discontinuity at the level of the septum or arterial wall. Both MRI/MDCT and ECHO (with or without contrast agent) can determine the shunt size, pressure gradient across the defect, and the onset of pulmonary hypertension.

Obstructions in CVM must be characterized by describing their presence, location and severity. They may be isolated or be part of complex malformations. Imaging can determine the gradient across the stenosed area and a gradient is used to select patients who may benefit from interventional therapeutic procedures.

Non-cyanogenic CVM with left-right shunt

Atrial septal defect – ASD represents 10% of congenital heart disease in children and 30% of congenital heart disease in adults older than 40 years [1,7-9]. There are several types of ASD, according to the location of the defect:

– ASD ostium secundum type, the most common [5] located in the middle portion of the interatrial septum, at the level of the oval fossa (fig 1);

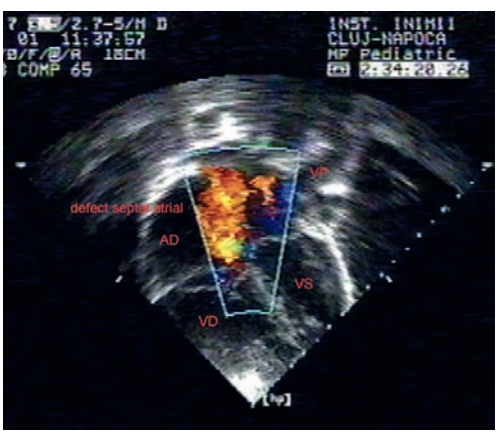


Fig 1. Color Doppler echocardiography. ASD type ostium secundum with left to right shunt across defect.

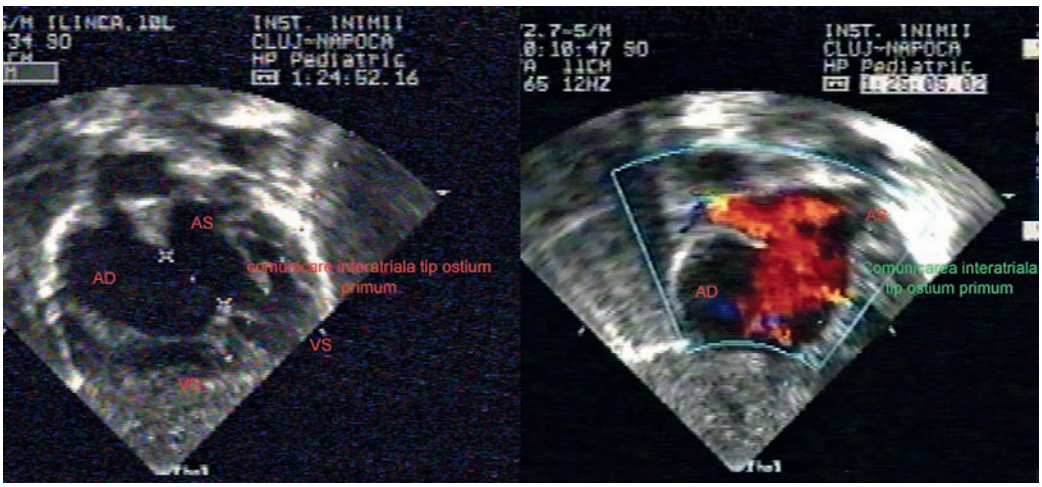


Fig 2. Echocardiography, subcostal window in B mode (left) and color Doppler (right), ASD ostium primum with left to right shunt

– ASD ostium primum type, second in frequency [8] low located, near the opening of the inferior vena cava into the right atrium (fig 2).

– ASD sinus venosus type, with high location, near the opening of the superior vena cava into the right atrium, being often associated with anomalous pulmonary venous return [8,10] (fig 3);

– ASD coronary sinus type produced by the dehiscence or absence of upper coronary sinus wall, thus communicating with the left atrium [1,3,6].

The reference examination is TTE [8], which allows in 2D B mode the location and the measurement of the opening and the assessment of indirect signs: dilated right heart cavities, dilated pulmonary artery trunk. Color Doppler obviates the blood flow in the defect.

ASD less than 3-5mm closes spontaneously mostly before 6 months. Postoperatively, the right ventricle returns to normal, if the atrial septal defect is closed under the age of 3 years [1].

TTE and TEE are limited in determining the pulmonary venous return, a situation where it may be necessary to turn to MDCT or MRI [3-5]. MDCT can also assess the appearance of large defects but it is limited in visualizing the small ones [5]. Bolus artifacts may alter the visualization of ASD. This is the reason to prefer a slower injection, obtaining a homogeneous opacification of the atrial cavities. Multiplanar reconstructions and 3D rendering provide important details in the pulmonary veins analysis (fig 4).

MRI offers both fine morphological analysis of ASD, with a sensitivity and specificity above 90% [8], and the most precise measurement of the shunt (pulmonary blood flow ratio - systemic flow Qp/Qs) [11-13]. It can also provide a reliable assessment of the anomalous pulmonary

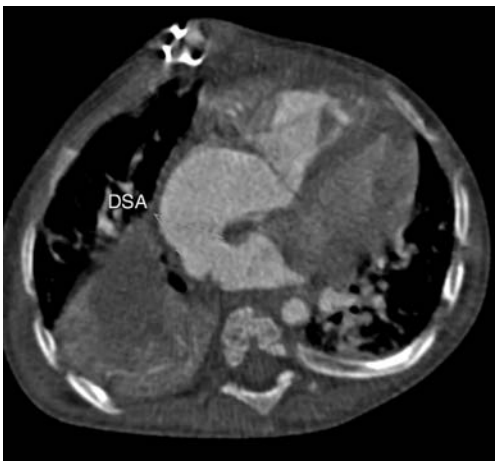


Fig 3. Angio MDCT axial section. ASD type inferior caval venous sinus.

venous return [2-5,8,16]. Synchronization of the acquisition with the electrocardiogram (ECG) is required (fig 5).

Ventricular septal defect. Inter ventricular communication represents approximately 20% of all cardiac malformations, being the second most common [1]. VSD sizes range from small and restrictive, to the form of a single ventricle (fig 6).

Inter ventricular septal defects are given by the lack of substance or mal-alignment of the various components, resulting in several types: membranous, perimembranous, of the inflow tract, of the outflow tract and muscular [3,8,17]. A large number of anomalies are associated with VSD: bicuspid aortic valve, coarctation of the aorta, mitral, aortic and tricuspid valve malformations, subvalvular aortic membrane and pulmonary atresia [1,3-5].

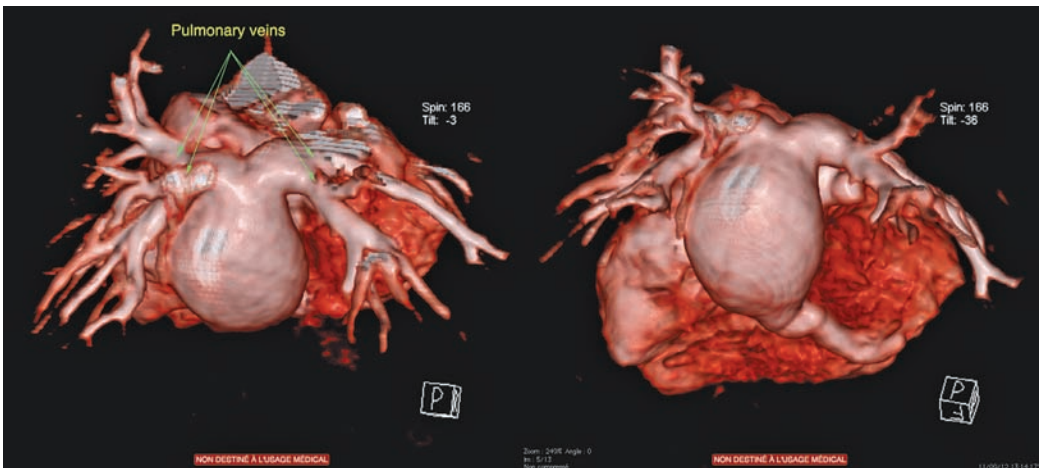


Fig 4. Angio MDCT, 3D VRT reconstruction. ASD type coronary sinus with total anomalous pulmonary venous return (RVPAT) – into common collector.

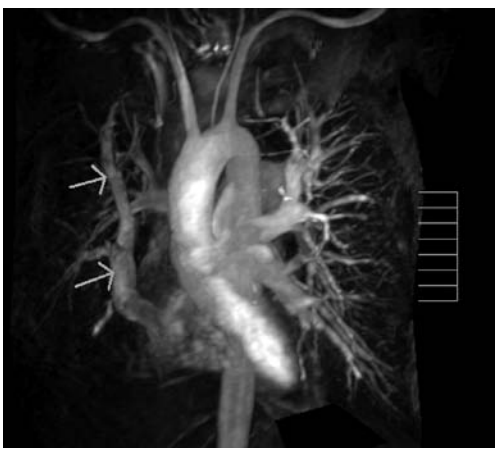


Fig 5. Angio MRI, 3D MIP reconstruction. ASD with right partial anomalous pulmonary venous return (RVPAP); the venous collector flows into inferior cava vein (VCI)

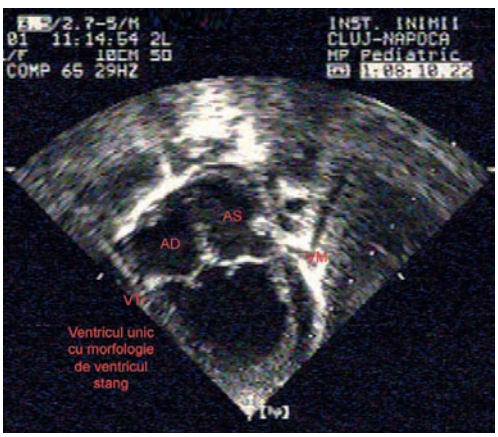


Fig 6. Echocardiography, B mode. Single ventricle.

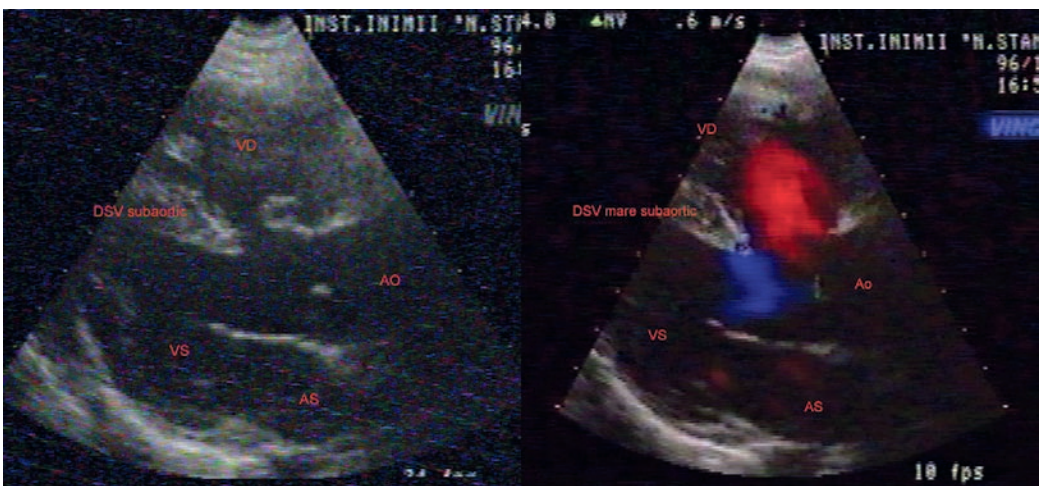


Fig 7. Echocardiography, B mode, parasternal long axis 5C (left), Doppler color (right). Large subaortic VSD.



Fig 8. Echocardiography B mode, subcostal abord short axis. Multiple muscular VSD.

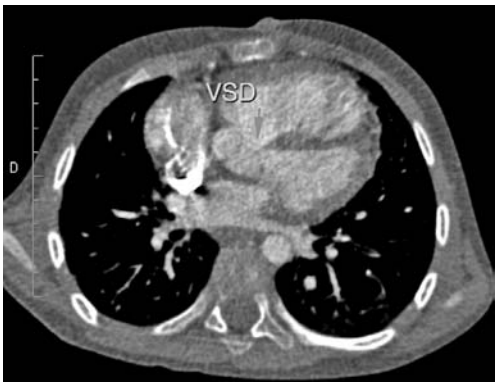


Fig 9. MDCT. Large VSD, mal – alignement of the aorta.

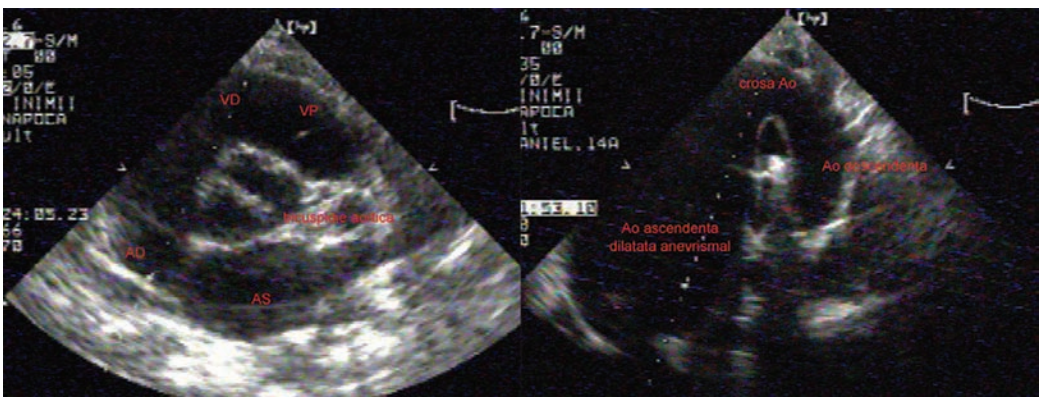


Fig 10. Echocardiography, B mode. Marfan syndrome: aortic bicuspid valve (left) and aneurysmal dilatation of the ascending aorta (right)

Echocardiography indicates the location, number and size of the VSD as well as mal-alignment, the relationship of the defect with the valves, and the tendinous cords [18,19]. (fig 7).

Muscular VSD, which are generally small, are difficult to assess with 2D ECHO (fig 8).

Doppler mode establishes the shunt direction, detects small defects and measures Qp/Qs ratio. Contrast enhanced ECHO can improve the detection of VSD especially by highlighting the right-to-left shunt in case of shunt reversal.

MDCT and MRI may visualize VSD, but are not indicated in their diagnosis when they are isolated, but only when associated with aortic coarctation or pulmonary atresia. These techniques are useful in the assessment of large defects and their postoperative follow-up. MRI is an excellent technique to measure the shunt flow rate as well as the right and left ventricular function [8,20-22] (fig 9).

Non-cyanogenic CVM without shunt.

Aortic stenosis comprises three types: subvalvular, valvular and supravalvular. The subvalvular type can be fixed or dynamic and may be due to the presence of a membrane or a muscle cushion from the interventricular septum, or it can take the form of a tunnel. It may be associated with obstructive or non obstructive hypertrophic cardiomyopathy or with VSD and aortic insufficiency. In valvular stenosis, the aortic valve has a "dome"-like appearance, it can be dysplastic, thickened or supple. The aortic valve is usually bicuspid (fig 10).

Echocardiography is limited mainly due to the reduced field of view. MDCT and MRI assess the valve, the stenosis, the morpho-functional characteristics of cardiac chambers and are the most accurate methods to measure the aortic diameter at all levels. They are also valuable in the characterization of potential complications (aneu-

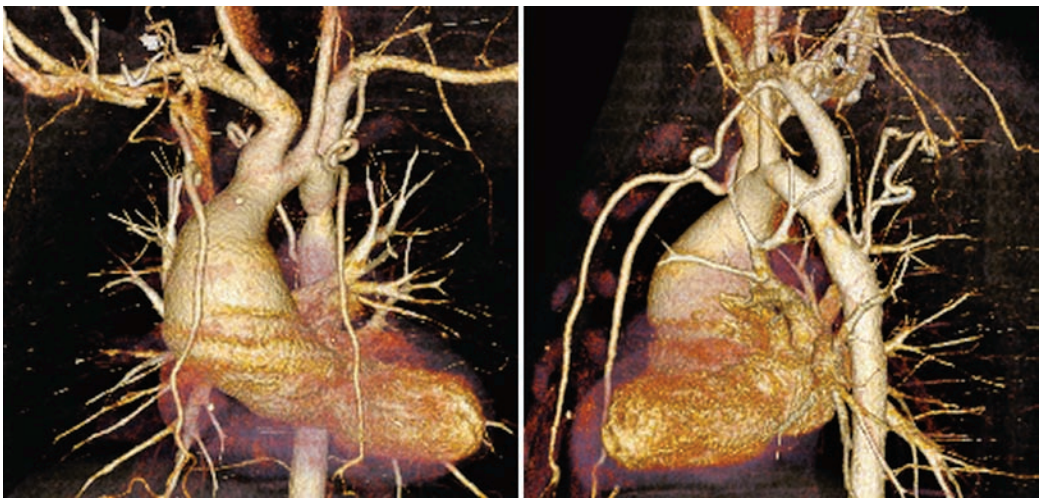


Fig 11. Angio MDCT. Marfan syndrome, aneurysmal dilatation of the ascending aorta, associated with aortic coarctation and collateral vessels.

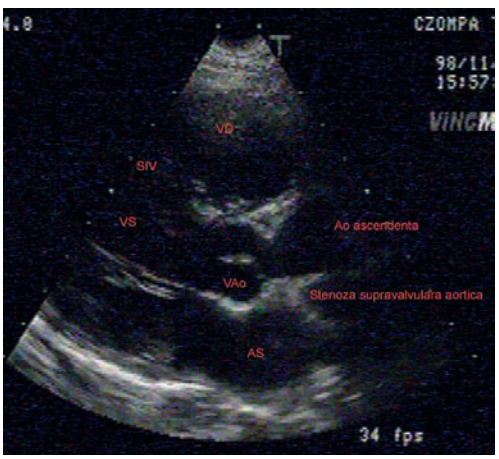


Fig 12. Echocardiography B mode. Congenital supravalvular aortic stenosis – «hourglass» appearance.

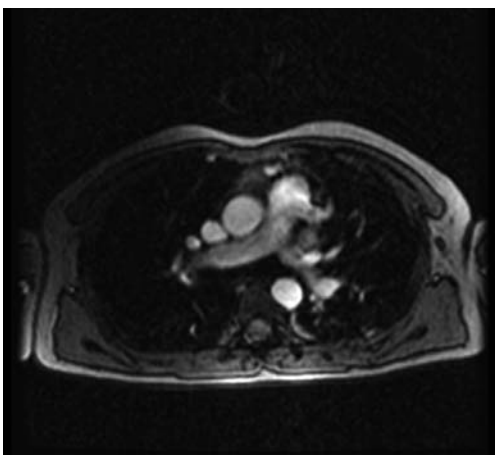


Fig 13. MRI cine SSFSE axial section. Anomalous origin of the left coronary artery from the pulmonary artery trunk.

rysm, dissection, rupture, thrombosis) and in evaluating supraaortic trunks and other emerging branches [4,5]. MDCT is the best method for quantification of valvular calcification, which hampers echocardiographic measurements (fig 11).

Supravalvular aortic stenosis may take the form of "hourglass". Echocardiography, MDCT and MRI angiography are very good diagnostic tools [24-27] (fig12). It is associated with aortic arch interruption, coarctation and hypoplastic aortic arch, abnormalities of the origin of the coronary arteries. Coronary anomalies are best characterized by MDCT and MRI (fig13).

The anomalies of the aortic arch (position, shape, number and the emerging supraaortic vessels) are best depicted with MDCT and MRI, both superior to ECHO [2,4,5]. The anomalies of the aortic arch and supraaortic branches are the result of primitive aortic arches abnormal development, through regression or abnormal persistence of one of the segments of the branchial arches. This results in vascular or vasculo - ligamentar rings around the trachea and esophagus, compressing neighbouring structures. They may have clinical respiratory (wheezing, dyspnea, respiratory distress) or digestive expression (feeding difficulties, regurgitation) or may be asymptomatic, discovered in adulthood [24,28]. Echocardiography is of limited value. Abnormal radiological appearance directs to MDCT angiography or MRI angiography [3-5,24,25,27].

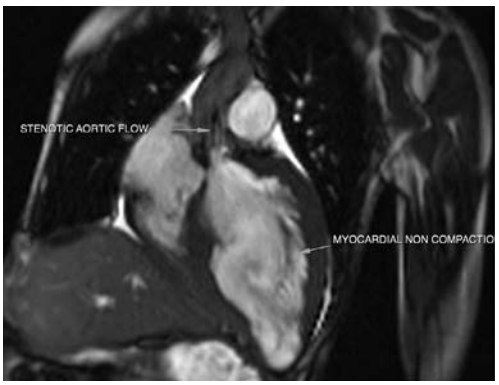


Fig 14. MRI. Congenital aortic valvular stenosis with myocardial noncompaction.



Fig 15. Angio MDCT. Aortic arch on the right side.



Fig 16. Angio MDCT 3D rendering. Double aortic arch with dominant right aortic arch.

The main types of anomalies are [24,28]:

– anomalies of the fourth branchial arch:

– normal aortic arch with aberrant right subclavian artery (arteria lusoria) – frequent anomaly, sometimes asymptomatic

– the aortic arch to the right with the aberrant left subclavian artery

– right aortic arch with mirror disposition, associated or not with Kommerell diverticula. This anomaly is frequently associated with congenital heart disease, most often with tetralogy of Fallot (fig 15)

– double aortic arch, with dominant right aortic arch, is the most frequent symptomatic vascular ring (fig 16)

– anomalies of the sixth branchial arch:

– pulmonary artery agenesis with absent or hypoplastic homolateral lung;

– retrotracheal left pulmonary artery – it is a rare, severe malformation, with symptoms obvious at birth in



Fig 17. Angio MDCT. Retrotracheal left pulmoary artery with tracheal compression.

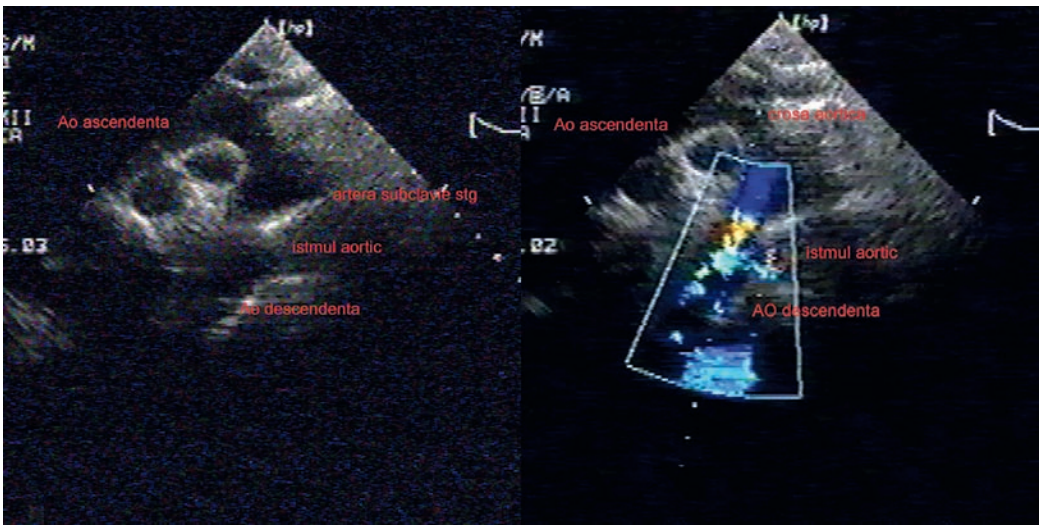


Fig 18. Transthoracic echocardiography, in B mode (left) and color Doppler. (right). Aortic coarctation.

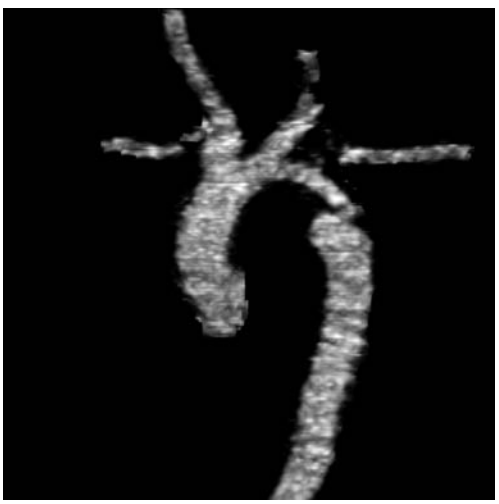


Fig 19. Angio MDCT. Hypoplastic aortic arch.



Fig 20. Angio MRI. Tight aortic coarctation.

which the left pulmonary artery emerges from the right pulmonary artery and has a retrotracheal course, compressing the trachea and right main bronchus [4,5,33] (fig 17).

Aortic coarctation. It is a common malformation, 4-10% of the total CVM [1] and it is part of the left heart obstructive disease. It can be located at the isthmic level, most commonly, preductal (associated with other malformations) or postductal [2,24].

The diagnosis is clinical and echocardiographic. The direct signs are: narrowed aortic isthmus just below the left subclavian emerging, associated or not with varying degrees of hypoplasia of the aortic arch and the presence of turbulent flow with high velocity immediately poststenotic (fig18).

The sign of severity is observing a continuous systolico-diastolic gradient in the aortic coarctation area, regardless of its value. Indirect signs are left ventricular hypertrophy and dilatation and turbulent flow through collaterals.

From a therapeutic standpoint, MDCT and MRI are very important both in children and, especially, in adults, because they can accurately depict the aortic morphology, they may indicate the existence and extent of collateral circulation and the presence of other associated abnormalities: VSD, bicuspid aortic valve, aortic stenosis, patent ductus arteriosus, presence of poststenotic aneurysmal dilatation [2-5,24-28] (fig 19, fig 20).

MRI is indicated in the diagnosis and postoperative follow-up [2,3,24] in the event of re-coarctation, to establish the indications for balloon and / or stent dilatation.

ECHO has many advantages, providing detailed anatomical and accurate functional data. It is an accessible, noninvasive and cost-effective method. ECHO allows monitoring of the natural evolution, intra-and postoperative follow-up, choosing the patients who require invasive investigation and interventional procedures. It contributes to the reduction of the indications of invasive procedures.

The indications for MDCT and MRI are well established, being complementary to ECHO and often replacing angiography. These new techniques have an important role in studying mainly the large vessels and their anomalies, as presented above.

Also, both methods play an important role in diagnosing CVM and their association within complex malformation syndromes with other abnormalities - skeletal, digestive, renal, broncho-pulmonary, cerebral, and also in the postoperative follow-up.

The main negative aspect of MDCT is irradiation of the patient. It is essential to use the scanner with moderation, with clear indications, to understand exactly what questions should be answered after the interpretation of images, when ECHO is of limited value. It is important to accommodate the examination protocol to the patient and to the pathology and to minimize, as much as possible, the irradiation.

MRI is the reference technique for exploring left and right ventricular function. MRI is able to measure the flow at any level (valves, vessels). Spatial resolution provided by MRI is inferior to MDCT. Also, as echocardiography, MRI is limited to viewing calcification and postoperatively, since sternoraphy or other materials are often the source of artifacts. In these situations, MDCT is the method of choice.

Another limitation of MRI is represented by the relatively long immobilization time required to complete the examination (30-45 min), often obtained only under general anaesthesia with controlled ventilation.

For young children or newborns, general anaesthesia is not required during MDCT, due to the performance in image acquisition speed.

Conclusions

Echocardiography is the technique of first choice in the diagnosis of CVM. It is complemented by MDCT and / or MRI to confirm or rule out anomalous pulmonary venous return in case of ASD and accurately assess cardiac function pre-and post surgery, in case of ASD or VSD. MDCT and MRI are important for complete and accurate diagnosis of the anomalies of the right heart chambers, the pulmonary venous and arterial tree, the emergence and course of the coronary arteries, coarctation of the aorta and aortic abnormalities.

The complementarity of ECHO with MDCT and MRI for obtaining morphological details and avoiding invasive procedures and the cooperation between the pediatrician, cardiologist, surgeon and radiologist, represent the key to the diagnosis and treatment in cardiovascular malformations.

Conflict of interest: none

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