Echocardiography and imaging investigation in congenital cardiovascular anomalies – competition or complementarity?
Part II: Cyanogenic cardiovascular malformations. Pictorial essay.

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Abstract
The diagnosis of cardiovascular malformations (CVM) is based on the echocardiographic evaluation. Multidetector computed tomography (MDCT) and magnetic resonance imaging (MRI) are performant, necessary techniques for the pre- and postoperative assessment of complex malformations, especially of cyanogenic malformations, in which anomalies of the right side of the heart and of the pulmonary circulation are involved and where echocardiography has a limited role. The complementarity of echocardiography with MDCT and MRI for the acquisition of the details necessary for an accurate therapeutic decision and for avoiding invasive exploration, as well as the close relationship between the radiologist and the clinician are crucial and all the more necessary in complex malformations.

Keywords: cyanogenic cardiovascular malformations, echocardiography, multidetector computed tomography, magnetic resonance imaging.

Cyanogenic cardiovascular malformations (CCVM) represent a large group of malformations, especially of the right side of the heart, which have in common the deoxygenation of the systemic arterial blood. The presence of cyanosis represents the key element in the diagnosis and it is more evident on the extremities, fingers, lips, ears, thus proving the circulatory character of the anomaly. Cyanosis is secondary to an obstacle associated with an upstream right-to-left shunt, the most typical example being that of the tetralogy of Fallot, or it can be secondary to anomalies of division or connection of the heart chambers and/or of connection between the cavities and the great vessels. CCVM may have a vital prognosis even in neonates and cause severe disabilities.

The prognosis of these malformations has greatly improved in the last years as a result of the progress that has been made in the area of diagnosis explorations as well as in that of the surgical techniques. Echocardiography is the examination of choice in these patients, but it has limitations in the exploration of the pulmonary arteries because of the air interpositions. In the postoperative phase the examination is also limited because of the surgical scar and the presence of sternal suture wires, all the more if it is an adult patient [1-5]. Multidetector computed tomography (MDCT) and magnetic resonance imaging (MRI) are performant alternatives in these situations, being especially useful in the preoperative assessment, but also in the postoperative follow-up, as they are able to give answers to specific issues [2-5]. MDCT has a superior spatial resolution compared to MRI, allowing the analysis of the lung parenchyma.
Tetralogy of Fallot is the most frequently encountered CCVM, accounting for 10% of the cardiovascular malformations [1]. It typically associates interventricular communication, the aorta displacement to the right, “overriding” the interventricular septum, infundibular pulmonary stenosis, and right ventricle hypertrophy. Actually, the main anomaly is represented by the anterior and cranial displacement of the infundibular septum, thus causing an obstruction of the right ventricular outflow at the level of the infundibulum [2]. The displacement of the infundibular septum results in a communication between ventricles due to a malalignment with the interventricular septum. The aorta is located above the ventricular septal defect. Right ventricle hypertrophy is secondary to the outflow obstruction.

Echocardiography reveals the interventricular septum discontinuity, the deviation of the aorta to the right, the mitral-aortic and aortic-tricuspid fibrous continuity; the ventricular septal defect can be large, even a common atrio-ventricular canal, pulmonary stenosis may be infundibular, valvular or it can be pulmonary atresia type (fig 1).

MDCT and MRI accurately provide important details such as the size of the ventricular septal defect, the degree of aortic overriding of the interventricular septum, the size of the right ventricular outflow tract, the diameter of the pulmonary artery, the diameter of the pulmonary branches and their continuity; the detection of coronary anomalies or other associated anomalies such as: persistence of left superior vena cava, persistence of the arterial canal (even double sometimes) or of the aortic-pulmonary collaterals, right aortic arch, etc. (fig 2, fig 3).

In practice the spectrum of this condition is much more complex: there is a minor form with a partial obstruction of the outflow tract (infundibulum), but with normal pulmonary arteries and a major form, extreme tetralogy of Fallot, which associates pulmonary atresia, with partial or complete absence of the pulmonary arteries in different segments of the lungs and systemic pulmonary vascularity through the persistent arterial canal and/or the major aortic-pulmonary collaterals [2,3]. The decisive element is represented mainly by the aspect of the pulmonary infundibulum and the pulmonary arteries [2,4,6,7].

**Fig 1.** Tetralogy of Fallot: a) B mode echocardiography: large interventricular communication (VSD) with right ventricular hypertrophy; b) Doppler echocardiography: VSD, aorta displaced to the right and subvalvular pulmonary stenosis.

**Fig 2.** Tetralogy of Fallot - MDCT: a) right ventricle hypertrophy and large ventricular septal defect; b) hypoplastic pulmonary arteries.
At birth there are two surgical options:
– complete surgical correction at 6-12 months
– in severe cases palliative surgery with Blalock Tas-sig shunt before the complete repair. This consists of an anastomosis with a GoreTex tube between the right or left subclavian artery and the homolateral pulmonary artery, through sternotomy or thoracotomy (fig 4). This way the pulmonary vascularity is maintained and the cyanosis is reduced, allowing the growth of the pulmonary arteries [7,8].

The complete repair consists of a sternotomy, closure of the ventricular septal defect using a patch and the widening of the infundibular-pulmonary tract. There are three possibilities:
1. transannular patch – is the most frequent surgical technique (it consists of an incision of the right ventricle and of the pulmonary valve ring, with a widening of the infundibulum and of the pulmonary artery trunk with a pericardial patch and a closure of the ventricular septal defect). The most important complications are: pulmonary insufficiency with right ventricle disfunction, pulmonary artery stenosis.
2. Rastelli procedure – it consists of an artificial conduit and valve between the right ventricle and the pulmonary trunk and the closure of the ventricular septal defect; this intervention is indicated in pulmonary atresia when a coronary branch crossing the anterior side of the infundibulum (this represents a contraindication for infundibulectomy). Possible complications: once the child grows the bridge must be changed as it becomes too short; calcification and stenosis of the graft; pulmonary insufficiency. MDCT is the best imaging method for the visualization of the coronary arteries (fig 5).
Fig 5. Tetralogy of Fallot – MDCT 3D reconstruction: coronary artery crossing the infundibulum.

Fig 6. Extreme tetralogy of Fallot with pulmonary atresia – MDCT: Prosthetic conduit between the right ventricle and the left pulmonary artery; anastomotic stenosis and distal left pulmonary artery stenosis.

Fig 7. Complication after tetralogy of Fallot repair – MDCT: a) Calcifications of the prosthetic tube; b) 3D VRT reconstruction: left pulmonary artery stenosis.

Fig 8. Extreme forme tetralogy of Fallot with pulmonary atresia. Prosthetic conduit between the right ventricle and the left pulmonary artery; anastomotic stenosis (red arrow) with infundibular dilatation (green arrow) - MDCT a) sagittal MPR reconstruction; b) 3D reconstruction.
In the case of the Blalock-Tussig palliative intervention, the most frequently encountered complications are:

– periprosthetic hemorrhagic suffusion that can be quickly and accurately visualized using MDCT; non enhanced CT acquisitions reveal periprosthetic hyperdensity and contrast media extravasation after injection [8];
– thrombosis of the prosthetic tube, which can be suspected on the x-ray as the pulmonary vascularity diminishes, but MDCT has the most important role as it shows the acute or chronic thrombosis after contrast media administration [3,9].
– pulmonary artery stenosis and/or subclavian artery stenosis, usually at the site of the anastomosis [3,9] (fig 6).

Tetralogy of Fallot has a long term favorable prognosis. There are a few complications that require subsequent surgical intervention: pulmonary insufficiency, aneurysmal distortion of the infundibulum, stenosis of the pulmonary arteries, residual valvular and/or infundibular stenosis, calcifications, stenosis or thrombosis of the pulmonary arteries, prosthetic tubes or infundibulum. MDCT and MRI are performant imaging techniques that allow the postoperative assessment in tetralogy of Fallot, as in some situations echocardiography has a limited value (fig 7).

Cardiac MRI is very useful in the evaluation of the patients with tetralogy of Fallot after surgery, allowing an objective appreciation of the pulmonary insufficiency and of the function and volume of the right ventricle, thus helping the decision for reintervention in order to place a pulmonary prosthetic [10,11].

The aneurysmal deformation of the infundibulum (fig 8) is caused by the pericardial patch which produces an infundibular dyskinesia, the backward flow determined by the associated pulmonary insufficiency favoring an important and disproportionate dilatation of the infundibulum. Diffuse infundibular dilatation of the whole pericardial patch must be distinguished from the infundibular pseudoaneurysm that occurs after a suture of the patch becomes dehiscent. The wall of the aneurysm is made of pericardium. In the false aneurysm, the dilatation is focal, the curvature of the aneurysm is shorter and connection angle with the heart is more acute. This situation represents a surgical emergency as there is a major risk for rupture. MDCT is the imaging method of choice in emergency situations.
**Pulmonary atresia with intact ventricular septum** is a very severe CCVM that is encountered only in newborns. Pulmonary atresia consists of a fusion of the two or three valvular flaps. The right ventricle is hypertrophic, has a small cavity, the tricuspid valve is abnormal, either stenotic or insufficient and the atrial septum is usually permeable. The pulmonary arteries are hypoplastic, but confluent. Maintaining a permeable foramen ovale and arterial canal is vital [12,13]. The role of MDCT and MRI is to visualize the pulmonary arteries and assess the postoperative shunts.

**Pulmonary atresia with ventricular septal defect** consists of a complete obstruction of the outflow trunk of the right ventricle, usually involving the valve, the proximal part of the pulmonary trunk and an interventricular communication. This cyanogenic congenital cardiopathy frequently associates anomalies of pulmonary angiogenesis. In severe forms the arterial branches are hypoplastic. A good knowledge of the pulmonary arterial tree is crucial for the surgical decision. MDCT and MRI have an essential role in the visualization of arterial pulmonary vascularity [2-4,15-19] (fig 9, fig 10).

There are several anatomopathological classifications of the pulmonary arterial tree, the most useful from a surgical perspective being the Barbero-Martial classification [14].

The surgical repair is complex, consisting of the reestablishing of the continuity between the right ventricle and the pulmonary arteries while achieving a pulmonary tree as close to normal as possible. Besides the analysis of the pulmonary arterial circulation the permeability of the arterial canal must be also evaluated. The arterial canal can emerge as usual from the aortic arch or, in situations when the aortic arch is deviated to the right, it can emerge from the brachiocephalic trunk, entering the right pulmonary artery.

The presence of a spontaneously permeable persistent arterial canal (fig 11) is a sign that the systemic to pulmonary collaterals are already developed. The collaterals can be [14]:

- direct aortopulmonary collaterals, emerging from the descending thoracic aorta especially its superior half, or from the subdiaphragmatic abdominal aorta. The collaterals can be from one and up to six for a lung and they are called major aortopulmonary collateral arteries – MAPCA; (fig 12, fig 13).
- indirect collaterals, emerging from the subclavian arteries or the brachiocephalic trunk;
- collaterals with bronchial origin in old patients;
- parieto-pleural collaterals.

When there are anomalies of the arterial tree with native or incomplete pulmonary arterial confluence, the technique of systemic-pulmonary collaterals focalization allows an improvement of the central pulmonary arteries caliber [20].

**Transposition of the great vessels (TGV)**

This type of anomaly involves a ventriculoarterial discrepancy: the aorta emerges from the right ventricle, while the pulmonary artery comes out of the left ventricle. There are two forms of malformation: the d-transposition (fig 14) with ventriculo arterial discordance and
the \textit{l-transposition} (fig 15), or the corrected transposition of the great vessels, with a double discordance, ventriculoarterial and atrioventricular. There are situations when the transposition is physiologically corrected, frequently in cases of dextrocardia or situs inversus.

Other anomalies may be associated with the TGV: Ebstein disease, other anomalies of the tricuspid valve, septal defects, and obstructions of the outflow trunk of the right or the left ventricle. Echocardiography has an important role in evaluating the aspect and the function of the tricuspid valve.

Complex forms with atrioventricular concordance and ventriculoarterial discordance evidence interventricular communication with the risk of developing a pulmonary stenosis, usually subvalvular, an aspect resembling tetralogy of Fallot.

In the isolated, double discordance transposition the malformation does not cause hemodynamic alterations. Yet the majority of cases demonstrate a large, interventricular communication, malformations of the right atrioventricular valve, pulmonary stenosis or atresia resulting in an appearance similar with that of the tetralogy of Fallot.

MDCT and MRI are useful in the pre- and postoperative evaluation of complex malformations, providing details regarding the connection of the great vessels, coronary arteries and the pulmonary vascular tree.

MDCT is a performant imaging technique useful in the appreciation of \textbf{pulmonary arterial hypertension}, including the Eisenmenger syndrome which develops...
secondary to the right-to-left reversal of a shunt, as part of a malformative syndrome. MDCT can accurately assess the size of the pulmonary arterial trunk and its branches; the changes occur in the pulmonary parenchyma and sometimes the compression of the dilated pulmonary trunk on the tracheobronchial tree.

**Conclusions**

Echocardiography allows the follow up of the natural evolution, the intra/postoperative assessment, and the long term follow up of patients with CCVM. Non invasive imaging techniques such as MDCT and MRI are complementary to echocardiography being very useful in completing the details of cardiovascular malformations, especially of complex CCVM. These techniques offer important informations in deciding the surgical approach.

**Conflict of interest:** none

**References**