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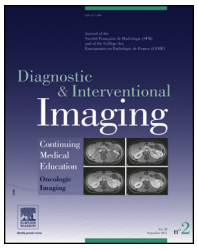

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CT and MR imaging in congenital cardiac malformations: Where do we come from and where are we going?

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Abstract The management of patients with congenital heart disease was profoundly changed firstly by the advent of pediatric and prenatal ultrasound and then more recently by cardiac magnetic resonance imaging (MRI) and computed tomography (CT) of the heart and great vessels. The improved life expectancy of these patients has brought about new medical and imaging requirements. MRI and CT are increasing second line techniques in this group of patients. This article summarizes the advantages and limitations of CT and MRI in some frequently encountered situations in children and adults followed up for congenital heart disease.

Where do we come from?

A long way away. Before the tremendous advances in diagnosis and medical and surgical treatment in pediatric cardiology, the situation of children described at the time as ''cardiac'' was not attractive. They had a high mortality rate, often at the end of a life peppered with complications. The life expectancy of patients even suffering from the less severe diseases remained short and imaging was restricted to radiography and invasive catheterization.

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Where are we now?

The situation has changed considerably, both in terms of diagnosis with the major input of echocardiography including prenatal investigations and in terms of treatment with the development of surgical techniques and endovascular treatments.

At present in France 6500 to 8000 children are born each year with cardiac malformations, and 85% of these reach adulthood [1]. These malformations, however, are still the main cause of death during the first month of life.

Antenatal diagnosis has revolutionized patient care. A typical example of this is transposition of the great vessels in which the antenatal diagnosis is often made in two stages: screening by a routine investigation followed by a reference antenatal echocardiogram allowing the birth to be organized with prompt care in a specialized center. As early as 1999, Bonnet et al. demonstrated the major impact of antenatal diagnosis on the children's survival [2]. These major advances have resulted in the emergence of a new population of adults suffering from congenital heart disease which our English-speaking colleagues refer to as ''GUCH'', ''grown-ups with congenital heart disease''.

The population in adult patients with congenital heart disease in France in 2013 was estimated to be 200,000 people. In a vast majority, this new population must be followed up by specialists (congenital disease cardiologists) and has created *ipso facto* a significant need for imaging. Nowadays, echocardiography has replaced chest radiography as the baseline investigation. The ''congenitalist'' will refer to a cardiovascular radiologist to supplement echocardiography and/or try to avoid invasive catheterization.

The choice between computed tomography (CT) and magnetic resonance imaging (MRI) is not always implicit and depends on the questions asked (and there is rarely only one of these). Occasionally, both investigations are performed, ideally at the same stage allowing the patient to have a single imaging session with a single preparation (insertion of venous access and sedation) and enabling the specialist radiologist to have a complete approach to the patient. In this situation, dual injections are avoided and the most suitable imaging investigation is chosen for enhancement. Also in this situation a report containing the findings from the two investigations is desirable.

The aim of this review was to guide the choice towards CT or MRI depending on the anatomy, age and clinical problem, reviewing the advantages and limitations of each of the investigations, based on a few typical examples.

The advantages of MRI

MRI should be used as a complement to echocardiography and not to replace it. Communication between the clinician and the imaging specialist is essential and the investigation request should be completed carefully and combined with all of the key documents (ECG, ultrasound, hemodynamic investigations, etc.). Direct contact is often useful in order to guide the technical protocol effectively. We should not forget that in congenital heart disease it is difficult to have a standard protocol and the physician needs to be close to the MR technician during the image acquisition. This involves basing the investigation on the questions asked and adapting the technique to the patient's abilities and cooperation. Real time interpretation is essential to the investigation.

The 2010 European guidelines propose that MRI should be used as a complement to echocardiography (adding to its results), or as an alternative to echocardiography when the latter offers insufficient information (for example a pauciechogenic patient) [3]. These guidelines emphasize the benefit of morphological and functional MRI investigation of the right ventricle (RV), pulmonary valve and arterial tract, aorta and by extension, all of the arterial and venous vessels in the mediastinum, together with its abilities to describe the myocardial tissue (fibrosis, iron or fat overload).

When the anatomical diagnosis is incomplete, a segmental approach is required and there is now consensus agreement on the Van Praagh model: this is beyond the purpose of this article and we would refer the reader to the excellent review by Lapierre et al. [4]. Briefly, the site of the viscera and atria are first established helped by the appearances of the bronchi and atria. Secondly the ventricular loop is determined by identifying the morphological features of both ventricles. Finally the vascular status is defined by examining the respective position of the aorta and the pulmonary artery. This approach may be carried out by MRI or CT or with a combination of these two techniques.

Bi-ventricular functional analysis is performed routinely and is rendered precise with a CINE b-FFE white blood image in apnea, whenever this is possible, usually performed along the short axis of the heart. If an apnea image is not taken, ''real time'' images are recorded, which offer less spatial resolution but allow approximate measurements to be made. Image processing is relatively non-automated in this disease context in view of the anatomical variations and the Simpson disc technique is to be preferred. It is essential to clearly identify the mitral and tricuspid planes on orthogonal sections (4 chamber views, vertical long axis of the LV and RV) in order not to include atrial sections in the ventricular surface areas measured (Fig. 1).

MRI is particularly useful to examine the RV, which is difficult to examine by echocardiography, and has a complex geometry. Morphological and functional examination of the RV is essential in patients who have undergone surgery for tetralogy of Fallot and also in patients with a systemic RV, for example after Mustard non-anatomical correction of transposition of the great vessels (D-TGV). When the congenital heart disease is complex it is recommended that anatomical views be taken along the conventional planes (transverse, sagittal and coronal).

In addition, MRI performs extremely well to investigate segments of the thoracic aorta, the examination of which is limited on echocardiography (specially the aortic arch and isthmus). Measurements of the aorta at the sinus of Valsalva and sinotubular junction are more accurate on MRI (or CT) than on echocardiography provided that they are taken along the true short axis of the vessel. Different measurement techniques can be used including black blood acquisitions, CINE (balanced FFE) or velocity encoding. Three-dimensional T1-weighted MR angiography (MRA) requires intravenous administration of a gadolinium chelate and it is not used routinely. MRA is particularly useful when stenosis or coarctation is present.