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Review

Multidetector computed tomography of congenital aortic abnormalities

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ABSTRACT

For many years invasive angiographic techniques have been considered as the gold standard for the assessment of large arterial abnormalities. However, the complexities and complications inherent to invasive imaging have meant that more recently non-invasive techniques such as echocardiography, Magnetic Resonance Imaging (MRI) and multidetector CT (MDCT) have been increasingly used in congenital cardiovascular disorders. In particular, MDCT has emerged as a fundamental tool for the diagnosis and pre-surgical work-up of aortic abnormalities due to its high spatial resolution, large area of coverage, and short scan time, and therefore is now one of the most widely used modalities for the detection of congenital abnormalities of the aorta. The purpose of this pictorial review is to review the spectrum of abnormalities of the aorta than can be reliably detected by MDCT both in infants and in adulthood. Abnormalities of the aortic root, ascending aorta, aortic arch, and descending aorta will be described separately.

1. Introduction

For many years invasive angiographic techniques have been considered the gold standard for the assessment of large arterial abnormalities. However, the complexities and complications inherent to invasive imaging have meant that more recently non-invasive techniques such as echocardiography, magnetic resonance imaging (MRI) and multidetector CT (MDCT) have been increasingly used in congenital cardiovascular disorders. In particular, MDCT has emerged as a fundamental tool for the diagnosis and pre-surgical work-up of aortic abnormalities due to its high spatial resolution, large area of coverage, and short scan time; it is now one of the most widely used modalities for the detection of congenital abnormalities of the aorta. The purpose of this pictorial review is to review the spectrum of abnormalities of the aorta that can be reliably detected by MDCT both in infants and in adulthood.

2. Abnormalities of the aortic root

2.1. Transposition of the great arteries (TGA)

Transposition of the great arteries (TGA) is characterised by a reversal of the main connections of the large thoracic vessels, such that the pulmonary artery arises from the left ventricle, and the ascending aorta arises from the right ventricle [1,2] (Fig. 1A). This is a cyanotic condition that is only compatible with life in the presence of an atrial or ventricular septal defect, or a patent ductus, in order to allow mixing of the systemic and pulmonary circulations; in these cases, mild cyanosis may be observed instead. On CT imaging, the aorta is often seen anteriorly (as the main vascular structure closest to the chest wall), and the pulmonary artery is noted to arise from the morphological left ventricle (Fig. 1B) [3]. For many years the Mustard procedure, which used artificial baffles to redirect blood appropriately, was used [4]. In contemporary practice the condition is corrected by means of the Jatene (arterial switch) procedure, ideally performed within the first two weeks of life [5,6] (Fig. 2).

In contrast, congenitally corrected transposition of the great arteries (ccTGA) is characterised not only by ventriculoarterial discordance, but also by atrioventricular discordance (Fig. 3). In such cases, the aorta again originates from the morphological right ventricle, which in such instances is seen to be positioned on the left. Conversely, the pulmonary artery originates from the morphological left ventricle, which is located on the right. As the pulmonary and systemic connections are preserved, this is an acyanotic condition, although it is often associated with other abnormalities (Fig. 4A, B). CT imaging shows the aorta to be located anterior and to the left of the pulmonary artery (Fig. 4A) [7].

It should be noted that both these conditions differ from the so-called "criss-cross heart", whereby the inlets of the two ventricles appear elongated and cross each other, thereby causing the apical portions

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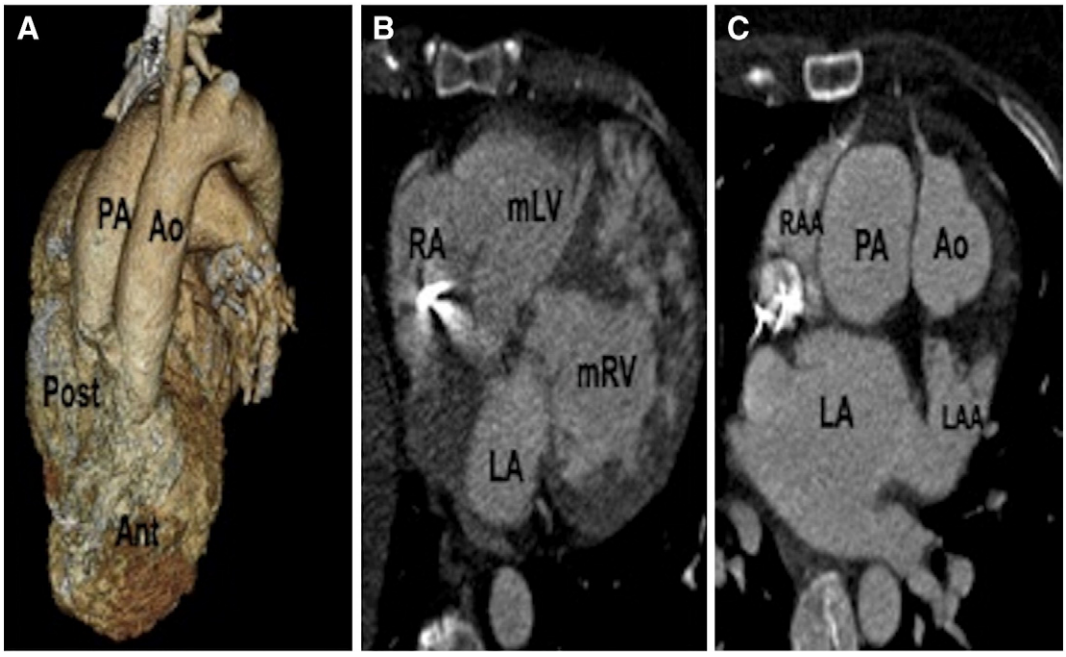

Fig. 1. A 30 year old male with a history of transposition of the great arteries (TGA) and Mustard procedure. (A) 3D volume rendered CT image and (B) sagittal CT image demonstrate ventriculoarterial discordance with the aorta (Ao) originating from the right ventricle (RV) and the pulmonary artery (PA) originating from the left ventricle (LV). (C) Axial CT image demonstrates RV hypertrophy (white arrowheads) and drainage of the pulmonary veins (PV) into the right atrium (RA). D. Coronal CT image demonstrates a metallic stent (black arrow) in the channel connecting the superior vena cava (SVC) to the left ventricle (LV).

of the ventricles to be situated opposite their expected locations [8] (Fig. 4C, D). In such cases, the ventricular outflow tracts and arterial connections are preserved.

2.2. Tetralogy of Fallot

Tetralogy of Fallot (ToF) is a common cyanotic heart defect, accounting for 3.5% of infants born with congenital heart disease and characterised by pulmonary infundibular stenosis (a narrowing of the right ventricular outflow tract), right ventricular hypertrophy, a ventricular septal defect, and the presence of an overriding aorta [9,10] (Fig. 5). The latter involves a biventricular connection of the aorta, which is situated above the VSD and connected to both the left and right ventricles (Fig. 5), however the degree of override is less than 50% (in contrast patients with double-outlet right ventricle are defined by a degree of override of greater than 50%).

Untreated, approximately 25% of ToF patients do not survive to one year, therefore early surgery is usually performed. For many years the condition was palliated by means of a Blalock–Taussig shunt, whereby an anastomosis is formed between the subclavian and pulmonary arteries to improve the flow of blood to the pulmonary circulation [11]. However in the modern era, total surgical correction is the preferred method of treatment [12,13].

2.3. Truncus arteriosus & aortopulmonary window

The truncus arteriosus forms in the embryo and normally divides into the aorta and pulmonary trunk. However, should it fail to do so then a persistent truncus arteriosus can present at birth with cyanosis, and the clinical syndrome rapidly deteriorates to heart failure [14] (Fig. 6). Two main classification systems are described: Collett–Edwards [15] and Van Praagh [16]; each system contains of four types based on the origin of the pulmonary arteries, however often the condition is associated with a right sided aortic arch as described above. Early treatment with surgical repair in the neonatal stage is advised, with evidence that surgical outcomes are improving [17].

An aortopulmonary (AP) window is also a form of communication between the ascending aorta and pulmonary trunk [18], however as opposed to truncus arteriosus this condition is characterised by the presence of two separate semilunar valves. The condition can be diagnosed by echocardiography, however MDCT can be useful in determining the location and size of the window (Fig. 6D, E, F).

2.4. Marfan syndrome

Marfan syndrome [19] is an autosomal dominant condition characterised by defects in the gene for fibrillin-1, a connective tissue protein [20,21]. It is characterised by a number of skeletal abnormalities (most characteristically tall height), however the majority of morbidity and mortality is caused by abnormalities of the cardiovascular system. The most severe of these is progressive dilatation of the aortic root, which often commences at an early age (Fig. 7). Progressive dilatation of the root can in turn lead to aortic incompetence, thus necessitating root and valve replacement surgery [22]. More recently a novel aortic root sleeve has been developed which is tailor-made to fit around the aortic arch and contain the dilatation of the aortic root [23], however this technique is in its early stages and long-term clinical outcomes are awaited. It must be remembered that other connective tissue conditions (e.g. Ehlers–Danlos syndrome, Loeys–Dietz syndrome) have also been associated with dilatation and aneurysms of the aortic root [24].