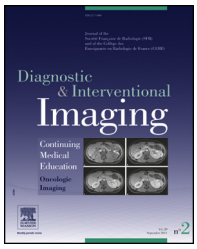
Diagnostic and Interventional Imaging (2016) **97**, 581—590



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# Multimodality imaging of aortic coarctation: From the fetus to the adolescent

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**KEYWORDS**
Coarctation;
Aorta;
Ultrasound;
CT;
MRI

**Abstract** Aortic coarctation is a local narrowing of the aortic lumen, which is located at the level of the isthmus in 95% of patients. Aortic coarctation accounts for 5 to 8% of all congenital heart diseases. It may have an acute presentation in the form of heart failure in the neonate or may be discovered incidentally in adult because of severe treatment-resistant hypertension. Ultrasound may reveal the presence of aortic coarctation during the antenatal period. In this situation, associated abnormalities should be investigated (including karyotype), because they influence prognosis and indicates whether or not the birth should occur in a center with pediatric cardiology expertise. Postnatally, ultrasound and chest radiography are the basic imaging work-up. Computed tomography is often the second line imaging investigation in infants and young children for whom magnetic resonance imaging fails to confirm the diagnosis. Magnetic resonance imaging with cardiac synchronization is the preferred imaging tool in the post-treatment period. Aortic coarctation may be treated surgically or by endovascular techniques. Potential complications should be searched for using ultrasound and magnetic resonance imaging.



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http://dx.doi.org/10.1016/j.diii.2016.03.006
2211-5684/

Aortic coarctation (AC) is defined as a local narrowing of the aortic lumen, which is located at the level of the isthmus in 95% of patients (i.e., between the origin of the left subclavian artery and the junction region between the end point of the aortic isthmus and the ductus arteriosus) (Fig. 1) [1]. AC accounts for 5—8% of all congenital heart diseases with a male-to-female ratio of 4/1 but its prevalence is undoubtedly underestimated. In this regard, Wren et al. reported that 54% of newborns with AC return home without any positive diagnosis [2].

AC is classified into two forms. The preductal form, which corresponds to an AC proximal to the ductus arteriosus (DA), has an early presentation. By contrast, the postductal form, which is associated with a closed ductus arteriosus, is usually discovered later in older children and young adults. Histologically, AC is accompanied by an abnormal presence of muscle fibers within the elastic fibers of the aorta [1]. These fibers arise from the DA and extend abnormally over the aorta to the insertion of the DA into the isthmus.

The goal of this review was to illustrate the major role of imaging in the assessment of AC.

## Classification

Two forms of AC have been described. In the preductal form, the AC is located proximal to a patent DA. Cardiac malformations are commonly associated. In the postductal form, the AC is associated with a closed ductus arteriosus. Atypical locations are possible, proximal to the isthmus or in the abdominal aorta. In the latter location, it is essential to search for extension to the renal arteries.

Classically, AC corresponds to a short stenosis, with a diaphragm allowing an excentric blood flow. Less commonly, AC can be a long and tubular stenosis. PseudoAC is defined by an excessive length of the aortic arch and the first portion of the descending aorta resulting in a folding or ''kinking'' effect next to the arterial ligament (Fig. 2).

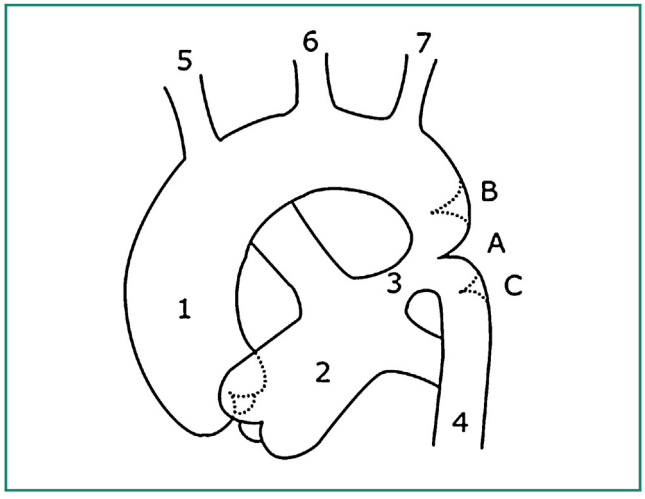


**Figure 1.** Diagram shows the various sites of aortic coarctation (1: ascending aorta; 2: main pulmonary artery; 3: ductus arteriosus; 4: descending thoracic aorta; 5: brachiocephalic arterial trunk; 6: left common carotid artery; 7: left subclavian artery; A: typical site; B: preductal form; C: postductal form).

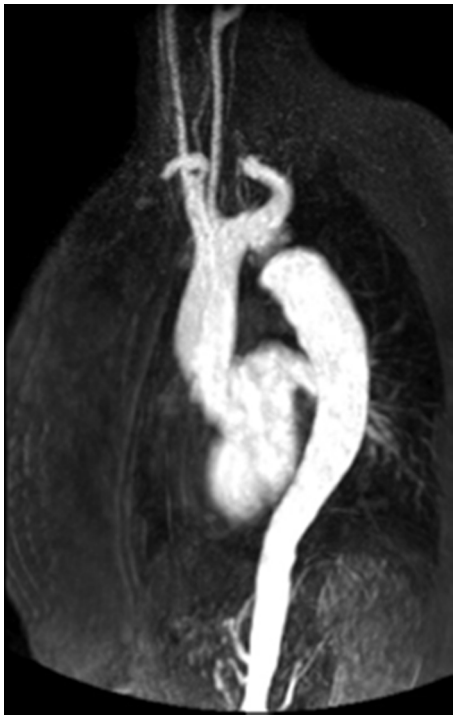

**Figure 2.** MRI angiography with maximum intensity projection reconstruction of a pseudo-coarctation due to excessive length of the aortic arch and the first part of the ascending aorta, resulting in a folding or *kinking* immediately next to the arterial ligament.

## Clinical findings

Clinically, AC can be diagnosed in the neonate presenting with acute heart failure or in adulthood as an incidental finding, usually because of severe hypertension and treatment resistance. Classically, pulses are abolished in the lower limbs in typical AC.

In the fetus, because the ductus is widely open, AC is ''asymptomatic'', or rather not expressed since not fully established, therefore difficult to diagnose on antenatal ultrasound [1].

Newborns with AC can present with:

- severe left cardiac failure with pulmonary edema and collapse;
- gradual heart failure with development of hypokinetic dilatation of the left ventricle;
- no clinical symptoms but asymmetrical pulses, upper limb hypertension or an abnormality on cardiac auscultation [3].

Seven to 13% of patients with AC have other cardiovascular malformations, including bicuspid aortic valve (60%), aortic arch hypoplasia (Fig. 3), mitral valve anomalies (stenosis, regurgitation or prolapse), in particular with Shone's syndrome (a combination of AC with a supravalvular mitral membrane, parachute mitral valve and sub-aortic valve stenosis), patent DA, atrial or ventricular septal defect (VSD), left superior vena cava, or transposition of the great vessels. Ten to 35% of patients with Turner syndrome have an AC (Fig. 4). Other related malformations are rarer: clubfoot, hypospadias, congenital cataract...