Diagnosis and evaluation of infant-type aortic coarctation: case series and review of imaging techniques

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Learning objectives

- Review of the main morphological anomalies seen in coarctation of the aorta (CoA), detectable with actual imaging technologies
- Literature review of the main imaging investigations used in evaluating CoA, and the main differences between them
- The role of different imaging techniques in the preop/postop evaluation of CoA
Background

Definition

Coarctation of the aorta is a hemodinamically significant, focal narrowing of the descending aorta, in the vicinity of the ligamentum arteriosum. [1]

Classification

In relation to the ligamentum arteriosum, the coarctation can be classified as either juxtaductal, pre- or postductal. [2]

There are different types of congenital obstructive lesions of the aortic arch, depending on the degree of transverse and/or longitudinal luminal stenosis:

1. Discrete coarctation of the aorta (shelf-like narrowing)

2. Tubular hypoplasia of the aortic arch

3. Combined hypoplasia and discrete coarctation (30% of cases) [3]


Type 1 obstruction is more commonly seen in the adults, while the more severe forms are diagnosed in infancy.

According to the location level of the obstruction with regard to the major vessels coming from the aortic arch, this anomaly can be classified in the following types:

A. Distal to the left subclavian artery

B. Between the left common carotid and subclavian artery

C. Between the innominate artery and the left common carotid artery [11]

Associated pathology

Aortic coarctation can be more frequently associated with the following pathology:

- Turner syndrome

- bicuspid aortic valve (in up to 70% of cases)[1]

- cerebral aneurysms [17]
- Shone complex [5]

The persistence of the Ductus Arteriosus (PDA) is almost always seen in infants having this pathology, and its role is to act as a shunt which can maintain a normal blood flow in the descending aorta. [11] This is why, in cases of severe hypoplasia of the aortic arch, the PDA is found to be very dilated, with its diameter comparable to that of the descending aorta.

While never seen in infants, aortic degenerative diseases (ascending aorta ectasia and aneurysm) and over-developed collateral circulation between the branches of ascending and descending aorta, respectively can be seen in older patients. [6,7]
Findings and procedure details

Although coarctation of the aorta has distinct clinical features (mainly a blood pressure gradient between the upper and lower extremities of at least 20 mmHg) which can lead to diagnosis, the morphological and functional characterization of this pathology can be done only by means of radiologic and imaging techniques. [8]

The signs which can be seen on a chest radiograph are mainly a projection of the degenerative modifications that take place later in the patients' life:

- "figure of 3" sign, represents the contour of the coarctation itself, with the upper portion being produced by the dilated ascending aorta and brachiocefalic trunk, and the lower portion, by the post-stenotic aortic dilatation

- rib notching, which can be seen in older patients (after early childhood), is a result of the erosion of the costal sulcus by the tortuous intercostal arteries. It can be seen from the fourth to the eighth ribs in the above cases [9]

Chest radiograph can still be considered useful as a 'first discovery' investigation in asymptomatic patients. [12]

Cardiac catheterization with manometry can be useful in detecting the morphology of the obstruction and estimating gradients estimating gradients. Together with transesophageal echocardiography, are also the main techniques available which can give insight information during minimally invasive surgery (balloon stenting). [8,17] Some disadvantages of the invasive angiography are: the risk of trauma to the vessels or the cardiac septum, the difficulty to cross the obstruction with the catheter from the distal approach, while also being impossible to gain access to the left side of the heart through the venous approach in the absence of a cardiac septal defect. [11]

Ultrasonography can give an accurate description of the morphology of the aortic arch obstruction, persistence of the ductus arteriosus (the existence of which must always be sought) and all associated cardiac anomalies. It can measure gradients through Doppler technology, but the results depend on the quality of the ultrasonic access, and also because it always tends to overestimate the values, so the results aren't useful for quantification, neither native, nor postoperatively. [8,10,11]

In neonates and infants, transesophageal echocardiography is difficult due to their narrow esophageal lumen, but it's described in the literature the use of an intracardiac probe as a transesophageal probe. The results were encouraging, the quality of the acquired data being comparable with that of MRI. [18]
In the literature, there are also described indirect echocardiographical predictors of the risk of an asymptomatic coarctation to become symptomatic (after the closure of the ductus arteriosus) [19]

Using fetal ultrasonography techniques (FINE - Fetal Intelligent Navigation Echocardiography), it can be 'raised the index of suspicion' for congenital heart diseases, including coarctation of the aorta. [22]

CT angiography can give a very [13] accurate assessment of the anatomy of the entire aorta, its branches, and also the orthogonal dimensions and relationship to other vessels, the presence and the precise dimensions of an existing hypoplasia of the arch, and also associated cardiac structural anomalies. [1] While the radiation exposure can't and shouldn't be underestimated, the wider availability when compared to MR devices, the data acquisition speed and the image detail quality, are all arguments in favour of using CTA in the diagnosis and management of this disease (surgical planning and follow-up). [15,16] This modality may obviate the need for invasive cardiac catheterization, thus exposing the patient to a much lower radiation dose. [19] Also, in neonates, it has been proven that cardiac CT can be performed free breathing and without sedation, with excellent results. [21]

Compared to MRI, the aortic arch is not as well seen by CT because it is imaged obliquely and reconstructed CT sagittal views have limited resolution. It is important to mention that measurements done with CT and MRI are not interchangeable, the differences between measurements being up to 9 mm, so, ideally, only serial measurements by the same method should be compared to minimize variability. [17]

MRA (Magnetic Resonance Angiography) can be used very successfully to demonstrate coarctation, and it's proving increasingly successful at assisting surgical planning. [11]

The MRI technique allows a more precise assessment of the severity of the obstruction and hemodynamics, including quantification of collateral flow, flow velocity, flow volume, and pressure gradients across the stenosis, using phase contrast. [1]

Multiple imaging planes are necessary to completely evaluate magnetic resonance imaging of coarctation, including oblique coronal and oblique parasagittal views. [14]

In patients who underwent corrective surgery using stents, MRI won't give quality information due to metal artifacts, and in this case CTA is the best option available. [17]

The following three cases will be presented as an example for the combined use of different imaging techniques with the purpose to assist in the diagnosis and further evaluation of the disease.

**Case 1**
Male patient, following a full term birth, weighting 4700g, was diagnosed in the neonatal period with aortic coarctation through echocardiography, which was requested after a routine clinical exam.

The relevant clinical exam results were a systolic BP of 150 mmHg, diastolic BP of 90 mmHg, and also weak distal pulse.

The aortic arch anomaly had an associated bicuspid aortic valve and also PDA, both of which were also diagnosed using echocardiography.

The coarctation was corrected through an invasive surgical technique, and the PDA was ligated.

In the following years, the patient received antihypertensive medication, in the more recent follow-up sessions accusing frontal headaches (beginning from 7 years old). A recoarctation is suspected, and, as a result, at 10 years old, a CTA examination is made, after echocardiography.

The echocardiography (done at 10 years old) report stated the following relevant findings:

- anomalous aortic valve with a median raphe between the right and the noncoronary cusp, and small aortic regurgitation
- small mitral regurgitation
- persistent atrial septal defect
- perimembranous interventricular septal defect significantly covered by a septal aneurism
- kinking of the aortic arch, in the vicinity of the left carotid artery
- ascending aorta diameter - 16.7 mm
- aortic arch diameter, proximal to the kinking - 12.1 mm
- aortic arch diameter at the level of kinking - 11.3 mm
- descending aorta diameter - 16.5 mm
- estimated maximal gradient at the level of obstruction - 23 mmHg, with a jet velocity averaging 2.55 m/s

The CTA report relevant findings were:

- the aortic arch, between the left carotid and subclavian artery emergences, suffers a 20% loss in diameter, with a length of 15-16 mm
- distal to the correction site, a slight post-stenotic dilatation can be seen, with diameters up to 21 mm, and with a length of 20 mm

- no collateral circulation over-development

- ascending aorta diameter - 22.1 mm

- proximal aortic arch - 11.4 mm

- distal aortic arch - 10.4 mm

- descending aorta diameter - 19.3 mm

The diagnosis of recoarctation was confirmed, and the patient underwent a corrective endovascular stenting procedure.

**Case 2**

Male patient, born at 36 weeks of gestation, after C-section, weighting 2000g at birth. The mother, who had a spontaneous abortion before, did not follow the mandatory pregnancy evaluation.

The patient was diagnosed after birth with DiGeorge syndrome. Tissue oxygen saturation was 74%.

The relevant results of the echocardiographic study were:

- ventricular septal defect (approximately 5 mm) with left-right shunt

- atrial septal defect (approximately 8 mm) with left-right shunt

- hypoplasia of the aortic arch, with possible interruption

- dilated PDA

- possible anomaly of the aortic valve

- possible transposition of the great vessels

- dilated right cardiac cavities

- mitral insufficiency 2nd degree

Due to the complexity of the associated malformations, and in the absence of suitable equipment and capabilities of the MRI devices available in the region, a CTA exam was requested.
The CTA report stated the following:

- Normal anatomical atrio-ventricular and ventriculo-arterial relationships
- Ventricular septal defect (5.4 mm in diameter)
- Atrial septal defect (2 mm in diameter)
- The aortic valve can’t be appreciated
- After the emergence of the brachiocephalic trunk, the aortic arch suffers a sudden decalibration, losing approximately 60% of its diameter in the proximal half and up to 75% distally, measuring 1.6 mm at this level. After the isthmic segment, the aorta recalibrates. Both the ascending aorta and the aortic arch have reduced densitometric values relative to the descending aorta, which has densitometric values comparable with the pulmonary artery.
- Dilated PDA (5.5 mm in diameter)

Case 3

Male patient, term birth by C-section, 3200g at birth, systolic murmur (3/6) and weak femoral pulses discovered at the routine clinical exam.

The relevant findings of the echocardiography report were:

- No atrial or ventricular septal defect
- PDA without significant dilatation
- Narrowing of the descending aorta, with a maximal gradient of 41 mmHg (maximal flow velocity 3.3 m/s) - minimal diameter is approximately 4.9 mm

The patient underwent a surgical correction (end-to-end anastomosis) with PDA ligation, and after approximately 4 months, he was reevaluated through echocardiography and CTA. Echocardiography results were inconclusive, estimating a residual gradient at the site of the obstruction of 23 mmHg.

The relevant CTA results were:

- Normal anatomical atrio-ventricular and ventriculo-arterial relationships
- The aortic arch suffers a decrease in its diameter compared with the ascending aorta (30%), continued distally, down to a diameter of 5.9 mm, with a slight increase at the junction with the descending aorta (7 mm). Distal from the left subclavian artery, the aorta
has a progressive narrowing in the isthmic portion, with a "funnel-like" appearance, down to a diameter of 3 mm.

- Very narrow PDA (1-2 mm in diameter)

- Emphysematous bulla located in the latero-basal segment of the lower right lobe

The patient underwent a corrective endovascular stenting procedure.
Fig. 1: Cardiac ultrasonography video loop depicting the aortic arch with coarctation in the vicinity of PDA

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Fig. 2: VRT image showing aortic arch hypoplasia

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Fig. 3: VRT video showing postoperative view of a corrective procedure for coarctation of the aorta

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**Fig. 4:** Candy cane view (MPR) - Case no. 1

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Fig. 5: VRT video loop showing the complex anomalies described at Case no. 2

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Fig. 6: Axial plane CTA scan - Case no. 2

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Fig. 7: Candy cane view (MPR)- Case no. 3

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Conclusion

- For the accurate highlight of complex morphological anomalies encountered in CoA, it's necessary to associate multiple imaging studies.

- Transesophageal echocardiography is able to accurately assess the morphological anomalies and their functional importance, with results comparable to that of MRI.

- Ultrasonography is useful in the diagnosis of the coarctaion of the aorta.

- MRI is becoming the imaging technique of choice for aortic coarctation evaluation and surgical planning.

- CT angiography is a fast and reliable technique for evaluating this anomaly, its associated morphological disorders, as well as its complications, but it lacks functional insight.
References


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