CASE PRESENTATION

Aortic coarctation in adults: the role of multimodality cardiac imaging. Series of case reports and review of literature

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Abstract: Coarctation of the aorta (CoA) is a relatively frequent congenital defect. Its natural evolution is marked by serious complications including aortic dissection, heart failure, coronary artery disease, infective endocarditis, or cerebral haemorrhages. Correction of CoA before complications arise is associated with a favourable long-term outcome. Timely diagnosis of CoA is therefore of utmost importance in the prognosis of these patients. Non-invasive imaging techniques, ranging from chest radiography to echocardiography, Cardiac Computed Tomography (CCT), and Cardiac Magnetic Resonance (CMR) have evolved to the extent where they can not only suggest but also precisely characterize the lesion and guide further management. We present a series of 3 case reports, highlighting the diagnostic approach and treatment for this pathology.

Keywords: coarctation, congenital, defect, multimodality, cardiac imaging.

INTRODUCTION

Coarctation of the aorta (CoA) is a relatively frequent congenital defect and represents 5-8% of congenital cardiac defects1. Its natural evolution is marked by serious complications including aortic dissection, heart failure, coronary artery disease, infective endocarditis, or cerebral haemorrhages. The estimated mortality in the case of untreated CoA is 90% by the age of 50, with 35 being the mean age of death2. On the other hand, correction of CoA before complications arise is associated with a favourable long-term outcome. Timely diagnosis of CoA is therefore of utmost importance in the prognosis of these patients. Non-invasive imaging techniques, ranging from chest radiography to echocardiography, Cardiac Computed Tomography (CCT), and Cardiac Magnetic Resonance (CMR) have evolved to the extent where they can not only suggest but also precisely characterize the lesion and guide further management. The current therapeutic options include open-heart surgery, balloon angioplasty, and endovascular stenting. We present a series of 3 case reports, highlighting the diagnostic approach and treatment for this pathology.

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CASE SERIES

Case 1
25 year-old male, with a past medical history of double ligation of patent ductus arteriosus at the age of 5, was admitted for poorly controlled hypertension despite optimal medical therapy. Clinical examination revealed a systolic interscapular murmur and a supine arm-to-leg noninvasive blood pressure gradient >20 mmHg (upper limbs systolic blood pressure - 155 mmHg, lower limbs systolic blood pressure - 80 mmHg). Echocardiography revealed hemodynamically significant post-ductal CoA with a peak Doppler systolic gradient of 95 mmHg (Figure 1A, 1B), a bicuspid aortic valve with normal opening, and mild regurgitation, dilatation of the aortic root, and ascending aorta. Cardiac computed tomography confirmed the presence of post-ductal CoA with an intrastenotic diameter of 12 mm, slight dilatation of the internal mammary arteries, and paths of collateral intercostal circulation (Figure 1C, 1D). Considering the severity of CoA in a patient with refractory hypertension, the patient underwent balloon angioplasty, and a covered stent was placed with no residual pressure gradient or significant stenosis (Figure 1E, Figure 1F) The first echocardiographic evaluation after correction revealed a marked reduc-
tion in the peak systolic gradient at the level of the dilated area (16 mmHg). Subsequent clinical monitoring revealed normal blood pressure values at 1 month and uncomplicated evolution at 5 years post-procedure. This case highlights the immediate and long-term benefits of catheter-based treatment in a hypertensive young patient with CoA, following surgical closure of a patent ductus arteriosus in childhood.

**Case 2**

38 year-old female with a past medical history of post-ductal CoA, surgically treated through Gore-tex patch aortoplasty at the age of 4, and grade III hypertension was referred to the hospital for cardiological evaluation. Clinical examination revealed a difference of 20 mmHg between upper and lower limbs systolic blood pressure. Echocardiography showed re-coarctation of the aorta with a peak Doppler systolic gradient of 42 mmHg (Figure 2B) at the level of the surgical repair, severely dilated left ventricle with moderate systolic dysfunction (Figure 2A) through diffuse wall hypokinesia (left ventricular ejection fraction - 35%), concentric hypertrophy, grade III diastolic dysfunction, and severe left atrium dilatation. Cardiac computed tomography with 3D reconstruction imaging confirmed the presence of CoA and allowed for its precise localization, extension, and measurement, as well as for the assessment of collateral circulation (Figure 2C, Figure 2 D). The coronary angiography was negative.

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**Figure 2.** Appearance of descending aorta in 2D suprasternal view (A) and high gradient measured at this level by continuous wave Doppler (B). MDCT visualisation of the aorta in the longitudinal MPR plane (C) and 3D volume rendering (D) showing aortic coarctation distal to the origin of left subclavian artery (yellow arrow). Angiographic view during stent deployment procedure: balloon post-dilatation (E) and final stent position (F). To note: the increase size of the internal mammary arteries.
Aortic coarctation in adults: the role of multimodality cardiac imaging

Carmen Beladan et al.

DISCUSSION

Aortic coarctation is considered to be part of a wider aortic pathology, with a large spectrum of morphologies, ranging from an isolated lesion to hypoplastic aortic arch segments. It is most often located in the thoracic aorta just after the ductal remnant and left subclavian artery, and in rare cases is located in the lower thoracic aorta, or abdominal aorta.

When deciding on the optimal therapeutic options, several factors come into play, including the anatomical severity of CoA, patient age, clinical presentation. The indications for correction of CoA or re-coarctation, as per the latest European guide for the management of adult congenital heart disease are outlined in Table 1.

It is generally recommended that corrective therapy of CoA be undertaken as soon as possible following diagnosis (preferably early during childhood) to reduce the long-term morbidity and improve the survival of these patients. However, in clinical practice, CoA is often diagnosed in late adulthood and poorly controlled systemic hypertension is a common clinical presentation in this setting.

Table 1. Recommendations for intervention in CoA or re-coarctation.

<table>
<thead>
<tr>
<th>Class. Level</th>
<th>Recommendations</th>
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<tr>
<td>I C</td>
<td>Repair of CoA or re-coarctation (either surgical or interventional) is indicated in hypertensive patients with an increased non-invasive gradient between upper and lower limbs confirmed invasively (peak-to-peak ≥20 mmHg).</td>
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<tr>
<td>IIa C</td>
<td>Stenting should be considered in hypertensive patients with &gt;50% narrowing relative to the aortic diameter at the level of the diaphragm even if invasive peak-to-peak gradient is &lt;20 mmHg.</td>
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<tr>
<td>IIa C</td>
<td>Stenting should be considered in normotensive patients with an increased non-invasive gradient confirmed invasively (peak-to-peak gradient of ≥20 mmHg).</td>
</tr>
<tr>
<td>IIb C</td>
<td>Stenting may be considered in normotensive patients with &gt;50% narrowing relative to the aortic diameter at the level of the diaphragm even if invasive peak-to-peak gradient is &lt;20 mmHg.</td>
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Data from 2020 ESC Guidelines for the management of adult congenital heart disease.

for significant focal coronary artery obstruction. Considering the presence of high-gradient aortic coarctation associated with poorly controlled hypertension and left ventricular dysfunction in a patient with re-coarctation who had been surgically treated in the past, catheter-based treatment was considered as the optimal therapeutic choice. The narrowed segment was identified and dilated (Figure 2E), and a stent was placed with no significant residual stenosis or gradient (Figure 2F). The post-procedural outcomes have been excellent, with a marked reduction in the peak systolic gradient at the level of the CoA (15 mmHg). One-year following the procedure, the patient was clinically well, with good blood pressure control and improved exercise tolerance. This case highlights the successful interventional treatment of aortic re-coarctation in an adult female patient, following surgical treatment in childhood.

Case 3

38 year-old male, with a past medical history of long-standing hypertension, was referred to clinic for cardiological evaluation. Clinical examination revealed a supine arm-to-leg noninvasive blood pressure gradient of >20 mm Hg and a grade II/VI systolic murmur, loudest in the right parasternal area. TTE raised the suspicion of CoA after identification of a systolic gradient with diastolic run-off across the descending aorta (Figure 3A, 3B). Cardiac computed tomography confirmed post-ductal CoA with a >50% narrowing of the aortic diameter compared to that at the level of the diaphragm (Figure 3C, 3D). Considering the anatomical severity and refractory hypertension, the patient was referred for interventional treatment. Procedurally, it was impossible to traverse the isthmic lesion via a femoral approach and therefore, a brachial approach was used for initial dilatation (Figure 3E). Once dilated, a covered stent was placed via a femoral approach, distal to the emergence of the left subclavian artery with good flow and no residual stenosis (Figure 3F). Following treatment, blood pressure levels normalized. The case highlights the successful catheter-based treatment of post-ductal CoA in an adult patient with a late diagnosis.

Diagnosis

Diagnosis of CoA usually starts with suggestive clinical features: upper limbs hypertension with concomitant lower limbs hypotension and a supine pressure gradient of >20 mmHg, a suprasternal thrill radiating to the back, decreased femoral pulse amplitude or radio-femoral delay, and palpable collaterals. Any of these clinical features will raise the clinical suspicion of CoA and lead to further evaluation.
Cardiovascular imaging is a central player in the clinical decision-making regarding patients with CoA. It provides crucial diagnostic information related to the anatomical characteristics and severity of CoA, hemodynamic significance, branching patterns, and length of involvement, as well as information on concomitant congenital heart lesions (bicuspid aortic valve, patent ductus arteriosus, ascending aortic aneurysm, subvalvular or supravalvular aortic stenosis, mitral valve abnormalities, Shone complex, etc.)^{3,4}. Imaging may also reveal extracardiac vascular anomalies including anomalous origin of the right subclavian artery, collateral arterial circulation, and intracerebral aneurysms^{5}. Moreover, it provides procedural guidance for operative or transcatheter intervention and post-procedural surveillance^{6}.

Chest radiography is a frequent initial diagnostic test, albeit of limited clinical utility. Specific signs may not always be present, but if they are, they can orientate further diagnostic methods. These signs include dilatation of the ascending aorta, notching of the inferior aspect of the 3rd-9th ribs due to development of collateral circulation (especially in older patients), and the „figure 3 sign“ given by the pre- and post-stenotic dilatation of the descending aorta, as was the case of the third patient presented above^{4} (Figure 4).

Given its ready availability and safety, as well as the possibility of assessing not only the site, structure, and
extent of CoA, but also the cardiac function, associated abnormalities, and collateral flow, transthoracic echocardiography is recommended as a first-line imaging method in the assessment of CoA. The suprasternal notch 2D echocardiographic view may provide the best imaging of CoA (Figure 1A). However, false images of aortic narrowing may result from a tangent sectioning of the vessel. Therefore, color Doppler flow mapping should be used to confirm the stenosis whereas continuous-wave Doppler is useful for estimating pressure gradient across CoA, albeit in the presence of extensive collaterals, these gradients are not reliable. The most reliable sign of significant CoA on the continuous wave Doppler is the presence of a diastolic anterograde flow due to a pressure gradient throughout the cardiac cycle (Figure 1A, 3B). Current European Guidelines do not recommend Doppler gradients for pre-operative quantification of CoA severity. In contrast, American Heart Association guidelines do define significant CoA as a mean systolic Doppler gradient of >20 mmHg or a mean systolic Doppler gradient of >10 mmHg in the presence of decreased LV systolic function, aortic regurgitation, or collateral flow. Diagnostic accuracy of CoA by 2D TTE imaging alone is reported to be 68%, however, the use of continuous-wave Doppler improved numbers to up to 90%. Visualization of CoA by TTE may however prove difficult at times due to poor acoustic window, long distance between the probe and isthmic region, and operator-dependence.

Transesophageal echocardiography (TEE) can provide precise measurements of the CoA diameter, with an accuracy reported to be similar to CMR in some studies. However, TEE Doppler examination is limited as the ultrasound beam is almost perpendicular to the direction of flow, thus providing inaccurate measurements. In clinical practice, TEE is not routinely used in patients with CoA given its limited added value.

Following the initial tests outlined above, CMR or CCT are the imaging techniques recommended by current guidelines to precisely evaluate the anatomy of the entire aorta and guide further management.

In addition to precise anatomic characterization, CMR can assess for the presence of collateral flow, evaluation of the myocardium, and associated anomalies (Table 2). Moreover, the combination of the narrowest CoA cross-sectional area and heart rate–corrected mean flow deceleration in the descending aorta obtained by contrast-enhanced 3D CMR angiography and phase-velocity cine CMR flow measurements, emerged as predictors of a catheterization gradient of ≥20 mmHg, with excellent sensitivity (95%), good specificity (82%), and an area under the ROC curve of 0.948.

Limitations of CMR include the exclusion of patients with certain metallic implants, nephrogenic systemic fibrosis occurring with Gadolinium administration and difficult image acquisition with signal artifacts in CoA corrected with stent implantation.

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**Table 2. Magnetic resonance imaging techniques used for the initial evaluation, pre-procedural assessment, and follow-up of patients with CoA**

<table>
<thead>
<tr>
<th>Technique</th>
<th>Description</th>
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<tbody>
<tr>
<td>Spin-echo CMR</td>
<td>Initial assessment of the location and degree of stenosis</td>
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<tr>
<td>Contrast-enhanced 3D CMR</td>
<td>Better visualization of the aorta in patients with repaired CoA</td>
</tr>
<tr>
<td>Phase-contrast, velocity-encoded cine CMR</td>
<td>Hemodynamic measurements - flow deceleration in descending aorta, pressure gradients Assessment of the smallest aortic cross-sectional area</td>
</tr>
<tr>
<td>4D flow CMR</td>
<td>Measurement of peak systolic pressure across CoA, wall-shear stress, and oscillatory shear index using computational fluid dynamics</td>
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Cardiac computed tomography represents a valuable tool in aortic imaging, with the advantages of wider availability and lower acquisition times. Dual-source CCT can evaluate the diameter of CoA with 100% accuracy\textsuperscript{13}. 3D volume rendering can aid in the characterization of the lesion, as reliably as during surgery or angiography. Moreover, using the multiplanar reconstruction technique, the ratio between the cross-sectional area of the aorta at the level of the coarctation relative to the aortic diameter at the diaphragm can be calculated and this can guide further management. A ratio of ≥50% indicates that angioplasty should be performed in hypertensive patients with CoA even if the invasive peak-to-peak gradient is <20 mmHg. Overall sensitivity and specificity of CCT for the diagnosis of CoA have been quoted to be 96.4% and 92.3% respectively\textsuperscript{4}. Disadvantages include the impossibility of providing hemodynamic information such as the pressure gradient or the degree of collateral circulation, the cumulated radiation dose, and exposure to contrast, however, these can be minimized with adequate use of CT protocols and state-of-the-art scanners. Optimal timing for scanning after contrast injection may be challenging in CCT, however, ECG-gating of image acquisition may significantly improve quality.

Catheter angiography remains the gold standard in evaluating the pressure gradient across CoA, obtaining high-resolution images of the aorta, describing the aortic geometry, and assessment of collateral flow. A peak-to-peak gradient of >20 mmHg in the absence of well-developed collaterals is indicative of significant CoA\textsuperscript{3}. Its invasive nature and exposure to radiation limit its use as a purely diagnostic method, however, it is currently preferred when correction by dilatation and stenting is considered. The incorporation of 3D CT and CMR models for pre-procedural planning and use in the catheterization lab may further aid optimal treatment\textsuperscript{14}.

**Treatment**

In deciding on the optimal treatment method for patients with CoA, non-invasive imaging provides essential information. For instance, in the presence of associated congenital heart and valve abnormalities, as evidenced by TTE, CCT, or CMR, surgical correction is preferred. Evidence of collateral circulation can be provided by TTE, CCT (Figure 5) or CMR and is useful in deciding whether to intervene or not. In patients with CoA, extensive collateral development, and mild hypertension, conservative therapy may be preferred\textsuperscript{15}. Additionally, if the patient proceeds with surgical treatment, which often involves clamping of the aorta, the absence of collateral circulation may mandate additional intraoperative steps to reduce the risk of spinal cord injury\textsuperscript{16}.

The combination of thorough pre-procedural non-invasive imaging is critical in enabling clear anatomic representations, especially in complex cases. A more cutting-edge technology is importing 3D rotational angiography datasets and overlaying the images over live 2D fluoroscopy\textsuperscript{17}, thus providing an accurate roadmap for device deployment.

Historically, the first interventional procedures consisted of simple balloon angioplasty. However, the usage of stents has been shown to result in a more effective reduction of the pressure gradient and a lower complication rate\textsuperscript{18}. Covered stents are now preferred due to the lower short and long-term complication rates\textsuperscript{19}. Biodegradable stents are currently being evaluated for the pediatric population especially, given that in time, stented aortic segments may exhi-

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**Figure 5.** Important collateral circulation with dilated internal mammary arteries visualized by echo (A) and by CCT (B) (red *). Low flow present at the level of superficial femoral artery (C). Decreased renal circulation with low flow measured in the arcuate arteries (D).
Organ characteristics, the chronic nature of the ver, this increases substantially beyond the age of 30-

Although several surgical techniques are available for children, graft interposition and bypass tube grafting are the only two feasible in adults. In cases with difficult anatomy, as evidenced by non-invasive imaging, ascending-to-descending aortic conduits may be preferred. Surgical risk in simple CoA is <1%, however, this increases substantially beyond the age of 30-40. Tissue characteristics, the chronic nature of the disease, co-morbidities, and collateral circulation development make this older age group a high-risk one.

Current guidelines recommend percutaneous angioplasty with stent placement as first-line therapy for both patients with native CoA and patients with recoarctation, in all cases where this is technically feasible. All three of our patients have undergone balloon angioplasty and stenting.

**Follow-up**

Despite good initial treatment results, an important number of patients go on to develop significant complications, a fact which highlights the need for regular follow-up using non-invasive imaging. The late cardiovascular complications post-repair include arterial hypertension, re-coarctation, complications related to the arterial wall (aneurysm, rupture, dissection, arteritis, fistula), and complications related to the stent (fracture, migration).

In 80% of patients who undergo repair during adult life, hypertension may persist despite complete correction of the aortic narrowing and long-term pharmacological blood pressure control may be required. This is correlated with increased LV mass, which can be easily assessed by TTE or CMR.

In 11-25% of patients reintervention for restenosis, as visualized by CMR or CCT, may be required. Aortic dissection or aneurysmal dilatation of the ascending aorta or at the intervention site can be evaluated by the above-mentioned imaging modalities too. Moreover, CMR has proved useful in the assessment of the adequacy of patch repair, angioplasty, or stent placement.

Some of the downsides of using CMR in the follow-up of these patients are that stent fracture cannot be reliably diagnosed and the usage of stainless-steel stents may result in sub-optimal image quality. On the other hand, CCT offers excellent in-stent imaging with the disadvantage of a cumulated radiation dose.

Current guidelines seem to agree on the main recommendations regarding follow-up and recommend yearly clinical evaluation. ESC recommends CCT (or preferably CMR) every 3-5 years, depending on the original pathology. Although the incidence of associated lesions such as intracranial aneurysms in CoA patients ranges from 2.5% to as high as 50%, European guidelines recommend routine screening for symptomatic patients only.

American guidelines recommend routine follow-up based on the Adult Congenital Heart Disease Anatomic and Physiologic classification system which uses both anatomical complexity and functional status in grading severity. Upper and lower extremity non-invasive blood pressure measurement is recommended in all patients. Based on severity, outpatient follow-up, including ECG, TTE, and exercise testing is recommended every 1-2 years, whereas CMR or CCT are recommended every 3-5 years.

**Conclusions**

Although most cases of patients with CoA are diagnosed during childhood, a significant proportion of patients are still diagnosed at an adult age. The most common reason for presenting is uncontrolled hypertension. In the diagnostic work-up of these patients, non-invasive blood pressure monitoring, chest X-ray, and TTE may provide clues to support the diagnosis, however precise anatomic evaluation by CMR or CCT is required before proceeding to angiography. Percutaneous dilatation and stent implantation represent the currently recommended first-line therapy in adults diagnosed with CoA, with surgical techniques being limited by the higher risk. Long-term outcomes are favourable; however, a significant proportion of patients may experience complications and thus regular follow-up (including CCT/CMR imaging) is required.

**Conflict of interest:** none declared.

**References**


