

COARCTATION OF THE AORTA

Definitions

Aortic arch obstruction may be due to:

Coarctation of the aorta (CoA) is a discrete obstructive lesion located just distal to the origin of the left subclavian artery where the fetal ductus arteriosus inserted into the aorta.

Tubular hypoplasia of some part of the aorta.

Aortic arch interruption.

Epidemiology

CoA is found in about 5-8% of patients with congenital heart disease, and is 2 to 5 times more common in males.¹ Coarctation of the aorta is primarily a non-familial or sporadic disease. However, it has been shown to coexist in families with left-ventricular outflow tract obstruction malformations such as supralvalvar aortic stenosis and bicuspid aortic valve that are markedly heritable. CoA is also present in 15% of patients with Turner syndrome who are at high risk of aortic dissection.² The most strongly implicated gene in non-syndromic CoA is *NOTCH1*. A missense mutation in the sarcomere gene MYH6 has also been associated with non-familial coarctation.³

Pathophysiology and Natural History

Obstruction of the aorta reduces flow the juxtaglomerular apparatus in the kidneys with resultant increase in vascular tone and intravascular volume. Thus, significant hypertension in the upper body occurs and collateral circulation develops in the form of **intracranial** (circle of Willis) and **intercostal artery aneurysms**. In adults a **bicuspid aortic valve** has been reported in 25 to 75% of patients with CoA. In children **VSD, PDA, and mitral valve abnormalities** are also common. Unoperated patients survive an average of 35 years with a 75% mortality noted by 46 years of age.⁴ Although the main cause of death in patients with corrected coarctation is coronary artery disease, coarctation itself does not predict for the development of coronary artery disease after adjustment for other risk factors such as aging, associated hypertension, hypercholesterolemia, and diabetes mellitus.⁵

Presentation

COA diagnosis and repair mostly occur in infancy and early childhood. However, it can be missed in childhood, leading to an initial diagnosis in adulthood, usually during evaluation for refractory hypertension.⁶ In early age CoA may lead to heart failure but the adult is usually asymptomatic. Rarely, headache, epistaxis, dizziness, palpitations and claudication

are reported. Diagnosis is suspected when hypertension is associated with diminished or absent femoral pulses. Patients with CoA have both ischemic and hemorrhagic strokes at significantly younger ages compared with the general population.⁷

Physical Examination

Every patient with systemic arterial hypertension should have the brachial and femoral pulses palpated simultaneously to assess timing and amplitude evaluation to search for the “brachial-femoral delay” of significant aortic coarctation (Tables 1 and 2). Supine bilateral arm (brachial artery) blood pressures and prone right or left supine leg (popliteal artery) blood pressures should be measured to search for *differential pressure*. There is differential systolic blood pressure (brachial-popliteal > 10 mmHg) and radial-femoral pulse delay unless significant AR coexists. The *diastolic* pressure is similar in arms and legs and therefore a widened pulse is felt in the arms.

Auscultation may reveal:

Interscapular systolic murmur

Widespread **crescendo-decrescendo systolic murmur** due to intercostal collaterals.

There may be also “**corckscrew**” **tortuosity** of retinal arteries.

An **ejection sound** and **midsystolic murmur** at the apex and base, respectively, can be heard in patients with a coexistent bicuspid aortic valve.

Investigations

Chest radiography may be normal. Rib notching (unilateral or bilateral) in 50% of cases.

The typical “figure-3” configuration of the aorta may also be seen in the PA projection.

ECG may show LVH.

Echocardiography is useful but CMR is the modality of choice, particularly for post-intervention surveillance. Every patient with coarctation (repaired or not) should have **CMR** or **CT** scan for complete evaluation of the thoracic aorta and intracranial vessels.

Detailed **fetal echocardiography** can stratify the risk for coarctation in fetuses with a suspected diagnosis. Prenatal detection rate of CoA may improve when additional criteria such as the presence of a coarctation shelf or hypoplastic arch are considered.⁸

Therapy

The natural history of unrepaired CoA includes the development of systemic hypertension.

Most untreated patients will die before 50 years of age and the condition should be diagnosed at a young age before hypertension develops.⁹ Early detection and treatment of CoA is associated with the best outcomes,¹⁰ although some patients will develop hypertension despite

repair. Patients with surgically repaired CoA who reach adolescence demonstrate very good long-term survival up to age 60 years. Long-term morbidity is common, however, related largely to aortic complications and late hypertension.¹¹

Surgical or endovascular repair indications

Indications for repair are presented in Tables 2-4 and Figures 1 and 2. Main indications are:

- CoA gradient >20 mm Hg
- Non-invasive pressure difference >20 mmHg between upper and lower limb with upper limb hypertension (>140/90 mmHg)
- Anatomic imaging evidence of significant coarctation with collaterals

Surgical repair

The surgical risk in simple CoA is <1% but it increases significantly beyond the age of 30–40 years and carries the risk of spinal cord injury. *Recoarctation, aneurysms or pseudoaneurysms* may occur in 10% of cases. When repair of CoA is performed between the ages of 20 and 40 years, the 25-year survival is 75%; in patients more than 40 years old the 15-year survival is only 50%.¹

Endovascular repair

Covered stent deployment is increasingly the treatment of choice for adults with comparable results with surgery.^{12 13} An uncovered stent has also been found safe and associated with persistent relief of aortic obstruction. Stent fracture and progression of fracture occurred but did not result in clinically important sequelae. Reintervention was needed in 20% of patients within 4 years, and related to early and late aortic wall injury and need for re-expansion of small-diameter stents.¹⁴ Stenting appears to offer lower acute complications compared with surgery patients or balloon angioplasty, but is more likely to require a planned reintervention.¹⁵ Percutaneous intervention (balloon or stent) is particularly recommended for recoarctation.

Treatment of hypertension

Systemic hypertension can occur after both surgical and endovascular repair and may be due to residual or recurrent coarctation, but even patients with a successful repair may develop hypertension. Risk factors for subsequent hypertension include an older age at the time of repair and higher blood pressure before the time of repair. The pathophysiology of hypertension that occurs after repair of coarctation of the aorta is not fully known.

Anatomical and functional changes in the arterial tree such as impaired elasticity and compliance and aortic stiffness may be involved. Post-repair surveillance for *hypertension, endocarditis or arteritis, and rupture of berry or thoracic aneurysms* is necessary. Even if the coarctation repair appears to be satisfactory, late postoperative thoracic aortic imaging should be performed to assess for aortic dilatation or aneurysm formation. Thus, lifelong follow-up is recommended for all patients with aortic coarctation (repaired or not). The

treatment of hypertension in coarctation of the aorta (CoA) is ideally managed by early surgical or transcatheter repair to reduce the risk of irreversible aortopathy and refractory hypertension. Medical therapy is the same as for essential hypertension, with beta blockers, angiotensin-converting enzyme inhibitors, and angiotensin receptor blockers as first-line agents.¹⁶

Pregnancy

It is allowed after repair. In unrepaired coarctation, residual significant hypertension or aortic aneurysms there is an increased risk of aortic rupture (Table 5).^{8,17}

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