Diagnosis, imaging and clinical management of aortic coarctation

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Received 11 January 2017
Revised 6 March 2017
Accepted 15 March 2017
Published Online First 4 April 2017

ABSTRACT

Coarctation of the aorta (CoA) is a well-known congenital heart disease (CHD) , which is often associated with several other cardiac and vascular anomalies, such as bicuspid aortic valve (BAV), ventricular septal defect, patent ductus arteriosus and aortic arch hypoplasia. Despite echocardiographic screening, prenatal diagnosis of C o A remains difficult. Most patients with CoA present in infancy with absent, delayed or reduced femoral pulses, a supine arm-leg blood pressure gradient (> 20 mm Hg), or a murmur due to rapid blood flow across the CoA or associated lesions (BAV). Transthoracic echocardiography is the primary imaging modality for suspected CoA. However, cardiac magnetic resonance imaging is the preferred advanced imaging modality for non-invasive diagnosis and follow-up of CoA. Adequate and timely diagnosis of CoA is crucial for good prognosis, as early treatment is associated with lower risks of long-term morbidity and mortality. Numerous surgical and transcatheter treatment strategies have been reported for CoA. Surgical resection is the treatment of choice in neonates, infants and young children. In older children (> 25 kg) and adults, transcatheter treatment is the treatment of choice. In the current era, patients with CoA continue to have a reduced life expectancy and an increased risk of cardiovascular sequelae later in life, despite adequate relief of the aortic stenosis. Intensive and adequate follow-up of the left ventricular function, valvular function, blood pressure and the anatomy of the heart and the aorta are , therefore, critical in the management of CoA. This review provides an overview of the current state-of-the-art clinical diagnosis, diagnostic imaging algori thms, treatment and follow-up of patients with CoA.

INTRODUCTION

Coarctation of the aorta (CoA) accounts for 5%–8% of all congenital heart disease (CHD) and occurs in 4 out of 1000 live births with a male predominance.[1 2] CoA can occur as a solitary lesion, but is often associated with other cardiovascular lesions, such as bicuspid aortic valve (BAV) (50%–75%), aortic arch hypoplasia, subaortic stenosis, mitral valve abnormalities, ventricular and atrial septal defects and patent ductus arteriosus (PDA).[3 4] The most important non-cardiac associated lesion is cerebral aneurysm in 2.5%–10% of patients with CoA.[5]

CoA is defined as a localised narrowing of the aortic lumen by a ridge, composed of medial wall thickening and infolding of aortic wall tissue.[4 6 7] CoA is also considered a general arteriopathy, given the often abnormal histology of the arterial wall adjacent to the site of coarctation and its association with long-term cardiovascular pathology. The narrowing is commonly located opposite to the insertion of the PDA (juxtaductal), but may also be located proximal (preductal) or distal (postductal) to the PDA.[4 8] The underlying pathophysiology of CoA is not fully understood. Three pathophysiological theories exist:

1. Abnormal embryogenetic development.
2. Reduced intrauterine blood flow through the aorta causing aortic underdevelopment.
3. Aberrant PDA tissue in the aortic wall, which constricts the aortic lumen at the isthmus, with postnatal PDA regression.[9]

Several genes have been implicated in CoA-aetiology, including the NOTCH1 gene, which plays an important role in cardiac development and vasculogenesis.[10] CoA is also associated with several syndromic phenotypes, such as Turner, PHACE, DiGeorge, Noonan and velocardiofacial syndromes.[11]

FOETAL DIAGNOSIS

Prenatal diagnosis of CoA reduces neonatal mortality and morbidity, prevents costly and risky transfers and reduces parental stress, as it allows for planned delivery in dedicated paediatric cardiac centres.[12 13] Prenatal ultrasound is a crucial primary screening modality.[14 15] Foetuses with an abnormal foetal heart or positive family risk factor(s) should be referred for foetal echocardiographic examination in a tertiary centre (figure 1).[14] Adequate examination of the foetal heart involves four recommended cardiac views: the three-chamber and four-chamber views, and the left ventricle and right ventricle outflow tract views.[13 15] Despite echocardiographic screening, prenatal diagnosis of CoA remains difficult.[16 17] CoA is the most commonly missed foetal CHD diagnosis, with less than one-third of the cases being detected at prenatal screening.[16 17] Concurrently, prenatal CoA screening is associated with a high number of false-positives, especially late in gestation.[12] Several echocardiographic markers may improve prenatal detection rates and reduce false-positives, including aortic arch z-scores, isthmus/ductal ratio, pulmonary artery/aorta ratio and the presence of continuous diastolic flow at the level of the isthmus.[16 18]

NEONATAL AND INFANTILE PRESENTATION

Most patients with CoA present in infancy (table 1).[19]

When the PDA closes after birth, severe CoA will lead to aortic obstruction with hypoperfusion of the lower body, renal dysfunction and metabolic acidosis.[4] Increased afterload on the left ventricle (LV) may lead to LV failure.[4] The presence of a PDA may obscure the diagnosis of CoA until it

To cite: Dijkema EJ, Leiner T, Grotenhuis HB. *Heart* 2017;**103**:1148–1155.

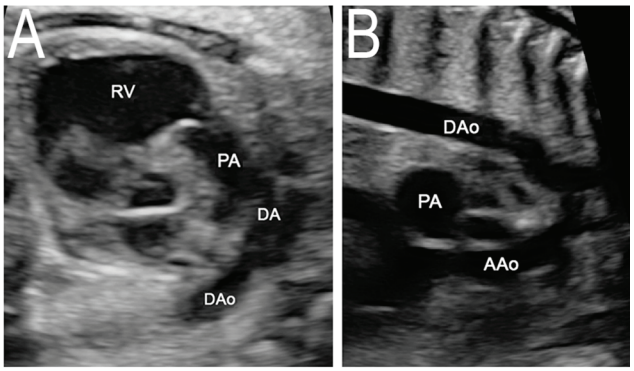

Figure 1 Foetal echocardiography in coarctation of the aorta: a shelf-like structure can be seen in (A) the ductal arch view and (B) two-dimensional sagittal view of the aortic arch. AAo, ascending aorta; DA, ductus arteriosus; DAo, descending aorta; PA, pulmonary artery; RV, right ventricle.

fully constricts, as aortic flow through the PDA may bypass the juxtaductal obstruction and supply output to the lower body.[8] Classical findings of CoA on clinical examination are absent, delayed or reduced femoral pulses, a supine arm-leg blood pressure gradient (>20 mm Hg) and a murmur due to rapid blood flow across the CoA or associated lesions like BAV.[8 20] Blood pressure readings should always include both arms, as an aberrant right subclavian artery (3%–4% of the patients with CoA) and diffuse aortic hypoplasia may obscure an arm-leg blood pressure difference or lead to a gradient over the transverse aortic arch, respectively.[21]

Neonates with clinically significant CoA should be hospitalised for continuous administration of prostaglandin E1, to keep the PDA open.[22] Patients with subtle clinical and/or diagnostic signs require close clinical and echocardiographic observation to determine whether PDA-closure results in clinically significant CoA.[22] Diagnosis of milder cases of CoA is important because hypertension and compensatory LV hypertrophy due to increased LV afterload may occur later in life.[6]

Table 1 Recognising CoA in patients of all ages

Age	Symptoms
Foetal	Ventricular disproportion (RV>LV) Great vessel disproportio Continuous diastolic flow at isthmus level Associated congenital heart disease (BAV, arch hypoplasia, subaortic stenosis, mitral valve abnormalities, VSD, ASD,PDA) Syndromic phenotypes (Turner, PHACE, DiGeorge, Noonan, velocardiofacial)
Neonatal and infantile	Collapse, acidosis Heart failure Systolic/ continuous murmur Weak or absent femoral pulses, radio-femoral pulse delay Supine arm-leg blood pressure gradient Necrotizing enterocolitis Renal failure Cardiomyopathy (rare)
Childhood, adolescent, adult	Systolic/ continuous murmur Weak or absent femoral pulses, radio-femoral delay Supine arm-leg blood pressure gradient Hypertension (headache, epistaxis, retinopathy) Reduced exercise capacity, exercise induced hypertension Leg fatigue and claudication Cold feet Left ventricular hypertrophy, arrhythmia and heart failure Infective endocarditis Aortic dissection, rupture Intracranial haemorrhage

BAV, bicuspid aortic valve; LV, left ventricle; RV, right ventricle; VSD, ventricular septal defect; ASD, atrial septal defect; PDA, patent ductus arteriosus.

PRESENTATION AFTER THE NEONATAL PERIOD

Post-infantile symptoms of CoA mainly depend on the location and degree of CoA, the extent of collateral circulation and presence of other CHD.[4] Symptoms may be similar as for the neonatal period, but can be absent in case of extensive collateral circulation.[9] Adult patients with CoA classically present with hypertension.[9] Intracranial haemorrhage secondary to intracranial aneurysms, LV hypertrophy and subsequent congestive heart failure can also be seen in adult patients with undiagnosed CoA.[4 9]

IMAGING

Chest radiography

Despite low sensitivity, chest radiography can raise suspicion for CoA.[23] A leftward convexity of the descending aorta with enlargement of the left subclavian artery can be seen in children and young adults with CoA, along with a figure-3 sign, formed by prestenotic and poststenotic dilatation of the descending aorta.[4] Rib notching of the posterior fourth to eigth ribs can be seen in older patients, related to collateral blood flow through the intercostal arteries.[4]

Transthoracic and transesophageal echocardiography

Transthoracic echocardiography (TTE) is the primary imaging modality for suspected CoA, given its ready availability, safety and capacity to provide haemodynamic parameters such as the CoA-gradient using Doppler (figure 2).[5] In addition, TTE can assess cardiac function and associated cardiac and valvular abnormalities.[4] Appropriate visualisation of the CoA-site can, however, be difficult due to a poor acoustic window and operator dependence.[7] TTE also has limited value in the evaluation of extracardiac structures and collateral circulation.[4 5 7] Although transesophageal echocardiography can provide accurate imaging of the aorta, it is seldom used due to its invasive nature and limited additional value.[23]

Computed tomography

CT provides high spatial resolution data of both intracardiac and extracardiac structures and allows for two-dimensional and three-dimensional reconstruction of relevant vascular anatomy.[5] Traditionally, the main disadvantage of CT has been the cumulative radiation dose from repeat examinations, especially in the paediatric population.[4 23] The introduction of multidetector CT with iterative reconstruction has significantly reduced radiation doses to values substantially below 1 mSv.[7 24] Furthermore, current state-of-the-art CT scanners can obtain the entire volumetric data acquisition in a single or several cardiac cycles, which minimises motion artefacts and can eliminate the requirement for breath-holding.[7 23] However, CT cannot provide haemodynamic information such as the pressure gradient across the CoA-site and the degree of collateral circulation, while the use of iodinated contrast media is associated with nephropathy.[4 5 23]

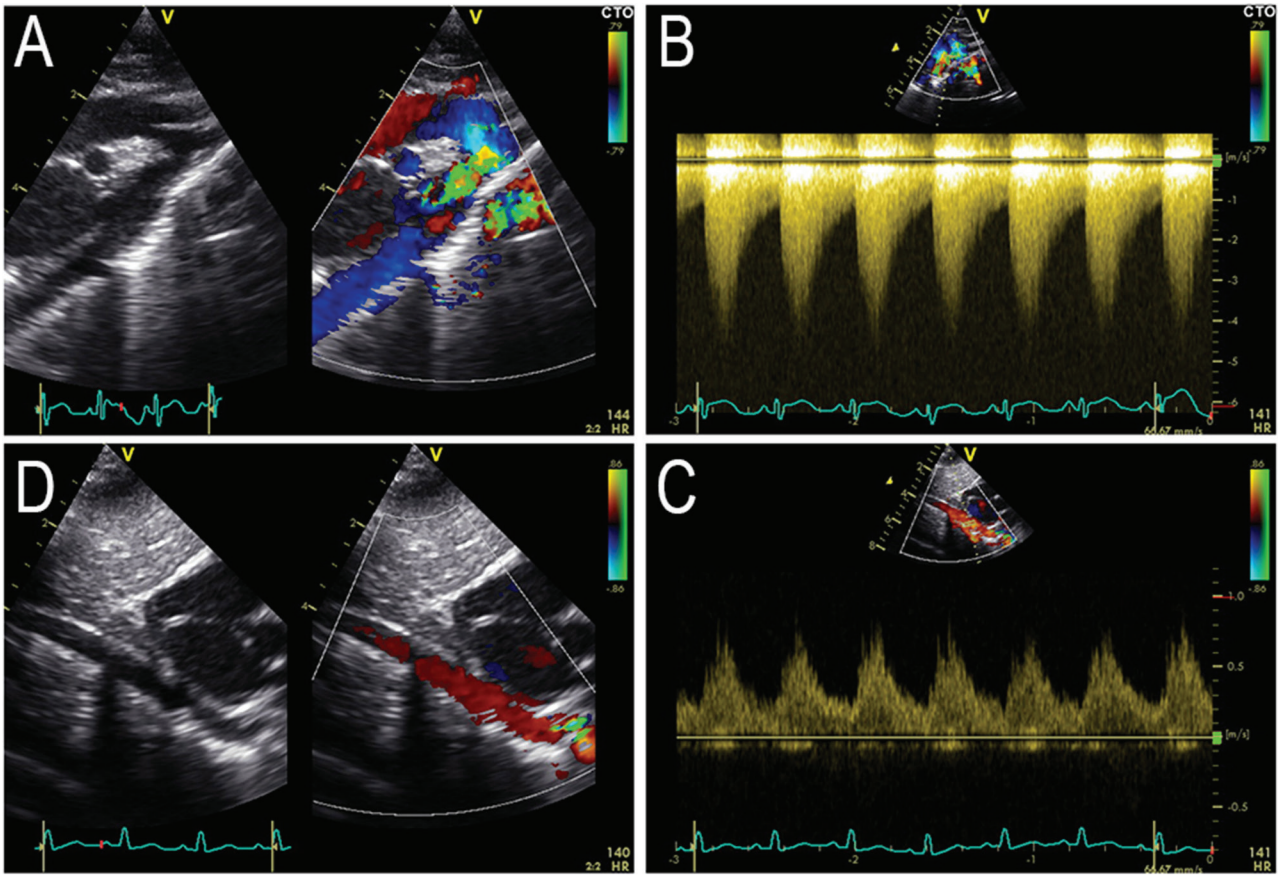

Figure 2 Transthoracic echocardiography with continuous Doppler showing high-velocity flow through the narrowed transverse arch and site of coarctation in the suprasternal notch view (A+B). Low-velocity, continuous flow was observed in the abdominal aorta (C+D).

Cardiac magnetic resonance imaging

Cardiac magnetic resonance (CMR) imaging is the preferred advanced imaging modality for non-invasive diagnosis and follow-up of CoA.[5 25] A major advantage of CMR is the lack of ionising radiation, making it ideally suitable for repeated imaging. Three-dimensional gadolinium enhanced CMR angiography provides excellent depiction of the aortic morphology and the location and degree of stenosis, as well as the extent of collateral vessel formation (figure 3).[25] Importantly, sole measurement of aortic dimensions may not be sufficient to assess the degree of CoA severity, as the haemodynamic effect of CoA is influenced by a complex interplay of aortic geometry, vessel wall mechanics, flow and ventricular function.[26 27] Therefore, more indirect measurements of stenosis—including the CoA-gradient—are often utilised.[27]

Cine imaging provides analysis of LV function and myocardial mass, whereas phase contrast flow analysis provides an estimated pressure gradient across the CoA and calculation of collateral flow.[23 25] Novel four-dimensional flow CMR allows for the assessment of flow velocities and pressure fields along the aorta, and the pressure gradient across a CoA, wall-shear-stress (WSS) and oscillatory shear index (OSI) can be calculated using computational fluid dynamics.[28 29] Despite these advantages, CMR is hampered by the long acquisition time and need for breath-holds during scanning, which restricts its use in smaller children and claustrophobic patients.[23] Sedation or general anaesthesia may then be necessary. In addition, metal stent implantation causes CMR artefacts, which limits adequate follow-up assessment.[23]

Catheter angiography

Catheter angiography is the gold standard for the assessment of the pressure gradient across the CoA and provides high-resolution images of the aorta and aortic geometry in 3D space (figure 4).[5] Catheter angiography also allows for accurate assessment of collateral circulation.[4] Catheter angiography is the traditional reference standard for CoA imaging, but the invasive nature and radiation burden are important drawbacks.[4 5] With the emergence of CMR and CT, CA is currently preferred when treatment options such as balloon angioplasty (BA) or stent insertion are considered.[4]

TREATMENT

If left untreated, the mean survival for patients with CoA is reduced to mid-adulthood due to long-term sequelae such as congestive heart failure, endocarditis, aortic rupture or cerebrovascular haemorrhage.[4 5] Life expectancy largely depends on the severity of the stenosis and the presence of associated anomalies.[5 8 9] Indications for CoA intervention are listed in box 1.[9 21]

Normotensive but significant CoA with substantial collateral flow should also be considered for treatment, as collateral flow reduces the perceived aortic gradient despite significant stenosis.[8 9]

Clinicians should aim for complete elimination of the aortic pressure gradient to preserve systolic and diastolic ventricular functions over time and prevent or at least reduce systemic hypertension.[21] Early intervention is preferred, as older age at the time of treatment is associated with increased risk of

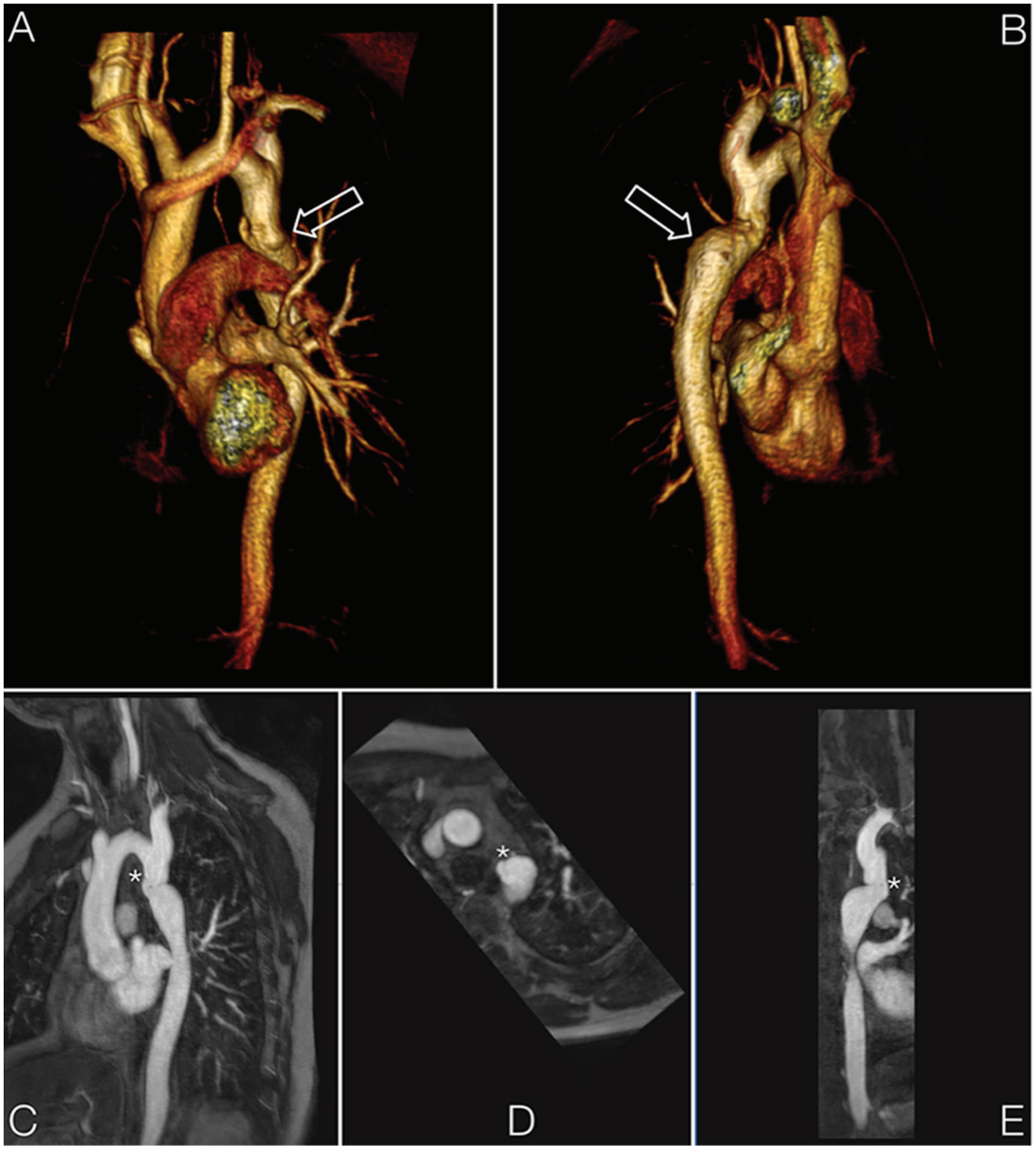

Figure 3 A 28-year-old man with known aortic coarctation. The examination was performed for follow-up reasons. Colour volume renderings in the left anterior oblique (A) and right posterior (B) views show the coarctation in the proximal descending aorta and the post-stenotic dilatation (arrows in A and B). Orthogonal reformations of the source data in the left anterior oblique (C), transverse (D) and right posterior (E) orientations show the aortic narrowing and kinking in more detail and reveal a small web in the inner aortic curvature (asterisks in C, D and E).

hypertension, aneurysm formation and mortality.[30 31] Timing of early intervention should, however, be weighed against the higher risk of restenosis and reintervention.[32 33]

Surgical repair

In neonates, infants and young children, surgical resection of the CoA-segment is the treatment of choice.[32] Several surgical techniques have been described. The surgical approach of choice is based on age and aortic arch anatomy.[9 34] Extended CoA resection with end-to-end anastomosis through a left thoracotomy is the preferred technique in neonates and children with discrete CoA.[32] In infants with long-segment CoA, subclavian-flap aortoplasty through a median sternotomy can be performed.[9] However, subclavian-flap aortoplasty has a higher chance of re-CoA and involves sacrificing the left subclavian artery (main arterial blood supply to the left arm).[9] CoA-resection and patch augmentation of the hypoplastic segment of the aortic arch are, therefore, preferred.

In adults, resection of the CoA-segment with graft interposition can be considered.[9] A bypass graft across the CoA-segment can also be considered in case of complex CoA, extensive collaterals or other significant cardiac abnormalities in need of repair.[9] Prosthetic-patch aortoplasty should be avoided due to high risk of aortic aneurysms and rupture.[9] Surgical mortality in CoA-intervention is rare (<1%).[35] Early postoperative morbidity includes paradoxical hypertension, injury to the recurrent laryngeal nerve (or other adjacent nerves) and subclavian steal syndrome (with subclavian patch angioplasty).[9] Although very rare, spinal cord

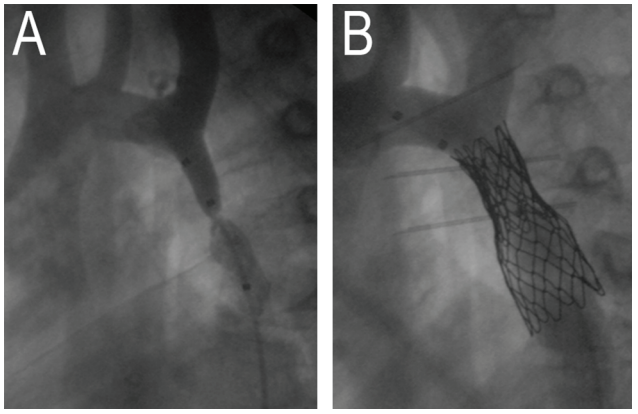

Figure 4 (A) Sub-atretic coarctation of the aorta in a 5-month-old girl with a gradient of 50 mm Hg across the sub-atretic segment. (B) The aortic arch after stent implantation with complete reduction of the aortic gradient.

ischaemia with paraplegia (due to prolonged clamping of the aorta) has been reported.[9]

Balloon angioplasty

BA is generally performed after 3–6 months of age because of high risk of re-CoA in younger patients due to reactive PDA tissue and the highly elastic properties of the aorta.[36] BA can be used in the neonatal period as palliative strategy in high-risk patients (ie, small or premature neonates) to stabilise their condition prior to definitive correction.[8 37] With BA, a catheter is brought up to the CoA-segment. The balloon size is chosen based on a maximum of 300% of the minimal aortic diameter.[21] To prevent aortic recoil, a therapeutic tear of the aortic wall into the media is created by overdilation of the vessel. After dilation, the restored blood pressure across the CoA-segment causes aortic wall remodelling, intended to create a lasting result.[8 21] Because of tearing of the aortic intima, BA predisposes for aortic dissection and rupture.[21] Long-term complications are recurrent stenosis and aneurysm formation.[21] In the past, a high incidence of 20% for aneurysm formation was reported for BA, but the risk is significantly reduced with technique-modifications, such as low-pressure, progressive or stepwise BA and smaller balloon sizes.[9 21] Re-CoA risks are relatively low for adults, but increase significantly with younger age at intervention (>50% for patients <1 year of age).[21]

Box 1 Indication for treatment of native coarctation of the aorta (CoA) and re-CoA

Indications for treatment:

- Supine non-invasive pressure gradient >20 mm Hg between upper and lower limbs
- Peak-to-peak coarctation gradient ≥20 mm Hg
- Peak to peak coarctation gradient <20 mm Hg with radiological evidence of significant coarctation with significant collateral flow
- Pathological blood pressure response during exercise
- Significant left ventricular hypertrophy
- Hypertension with ≥50% aortic narrowing relative to the aortic diameter at the level of the diaphragm
- Upper limb hypertension

Stent placement

Endovascular stent placement provides a solution to elastic tissue-recoil after BA.[9] In older children (>25 kg) and adults, transcatheter treatment is the treatment of choice because of good results and the less invasive nature of this technique.[9] Endovascular stenting is preferred over BA alone, with lower risk of restenosis and aneurysm formation.[38] Stent implantation in young children remains controversial due to the need for frequent redilation to accommodate the growing aorta, lack of available redilatable stents, high incidence of intimal proliferation and restenosis and risk of poststent aneurysms.[37] Recently, the usage of growth stents and biodegradable stents in children has been described,[39 40] although long-term results of these new techniques have not been reported yet.[8]

With endovascular stenting, a stent is dilated with inflation of a balloon until the stent is anchored in the aorta and the stenosis is sufficiently relieved (figure 4). Rapid ventricular pacing can be used to lower the stroke volume and facilitate proper stent placement.[21] Stent placement provides a more sustainable relief of the CoA-gradient, with more even distribution of forces across the vessel wall.[9] The introduction of covered stents—as an alternative to bare-metal stents—has further reduced the risk of aneurysm formation.[30] Despite these advantages, stent insertion is technically more complicated than BA and requires a bigger catheterisation sheath, resulting in higher risks of complications at the insertion site.[21] Rare complications of stent placement include stent migration and/or embolisation, occlusion of aortic side branches with covered stents and aortic dissection and rupture, associated with cerebrovascular accidents in very rare cases.[21] Long-term complications include restenosis, stent fracture and aneurysm formation.[30]

FOLLOW-UP

Despite excellent long-term survival after CoA-repair, patients have a reduced life expectancy with increased risk of morbidity.[33 41] Long-term complications occur despite adequate and timely repair, warranting life-long follow-up (table 2). Annual blood pressure evaluation (four limbs) and regular imaging of the heart and aorta are recommended.[32 42]

Re-stenosis and reintervention

Cardiovascular reinterventions are common (25%) among patients with CoA.[43] Indications for reintervention are re-CoA, aortic valve dysfunction and other cardiac malformations (ie, VSD).[43] Recoarctation is seen in 4%–14% of the patients,

Table 2 Long-term complications of CoA

Long-term complications	
Local	Recoarctation, aneurysm, dissection, rupture, fistulae, endocarditis
Ascending aorta	Aneurysm, dissection, rupture, sinus of Valsalva fistula
Aortic valve	(Bicuspid valve), stenosis, regurgitation
Left ventricle	Hypertrophy, dilation, systolic dysfunction and/or diastolic dysfunction, heart failure, sudden cardiac death
Coronaries	Premature atherosclerosis, ischaemic heart disease
Cerebral	Berry aneurysms, intracranial bleeds, atherosclerosis, stroke
Systemic	Hypertension, reduced exercise capacity

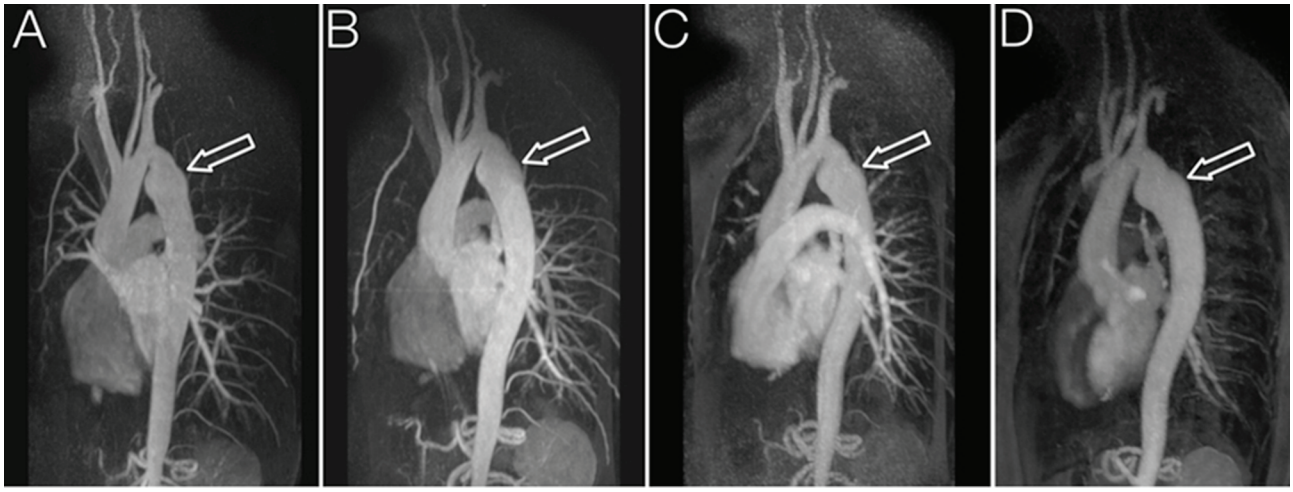

Figure 5 A 26-year-old man with known aortic coarctation imaged four times over a 10-year period shows mildly progressive dilatation of the poststenotic part of the proximal descending aorta over time (arrows). The study in A was obtained in 2007; the study in B was obtained in 2010; the study in C was obtained in 2013 and the most recent follow-up examination was performed in 2016 (D).

caused by residual ductal tissue, scarring, unrelieved arch hypoplasia and intimal hyperplasia after stenting.[34 43] Repair before 1 year of age carries a greater risk of re-CoA.[32] Re-interventions occur more frequently after stent placement or BA, but these reinterventions are often anticipated with growth of the child.[30] Unplanned reintervention rates are similar for surgical and transcatheter treatments.[8] Indications for treatment are similar as for native CoA.[9 38] More than half of the patients with CoA suffer from a BAV, which is associated with a high risk of aortic valve repair or replacement surgery during long-term follow-up.[3] Indeed, half of all reinterventions in patients with CoA are due to aortic valvulopathy, so assessment of aortic valve function should be part of follow-up.[3]

Generalised vasculopathy and aneurysm formation

The prestenotic arch and conduit arteries of patients with CoA show abnormal vascular compliance due to reduced smooth muscle tissue and increased collagen, leading to increased intimal-media thickness.[19 44] The prestenotic arch also demonstrates endothelial dysfunction, possibly related to altered aortic flow dynamics.[28] Associated increased WSS and decreased OSI result in higher atherosclerotic susceptibility.[45] Abnormal compliance and endothelial dysfunction can persist after CoA-repair and are associated with generalised arterial vasculopathy later in life.[46] Vascular complications include aortic aneurysm, hypertension, aortic root dilation, aortic and mitral valve insufficiency, premature coronary artery disease and cerebrovascular disease (figure 5).[32 42] Aortic aneurysm is defined as >150% dilation of the repair site in relation to the aorta at the level of the diaphragm and occurs with an incidence of 13%.[43] Aneurysms are mostly seen after patch angioplasty and less often after BA or subclavian flap surgery.[38 43]

Hypertension

Chronic hypertension remains present in 35%–68% of the patients with CoA, even in the presence of an anatomically satisfactory repair.[31 33 42] Furthermore, exercise-induced hypertension occurs in over one-third of the normotensive patients.[44] Prevalence of systemic hypertension is significantly lower in patients treated in the neonatal period or infancy and in patients who underwent CoA resection with end-to-end anastomosis.[32 33] The exact pathophysiology of late-onset hypertension after CoA-repair remains to be elucidated. Reduced aortic compliance, abnormal baroreceptor function and altered WSS dynamics may play a role.[31] Hypertension is also associated with an abnormal geometry of the aortic arch ('Gothic arch' configuration) when the arch is sharply angulated in the sagittal plane.[33] Mild residual morphological obstruction and a disturbed renin–angiotensin–aldosterone system appear to play no major role in late-onset hypertension.[19] Post-treatment hypertension is a risk factor for premature death and requires aggressive treatment.[31] Follow-up should include 24 hours of blood pressure measurement and exercise testing, as exercise-induced hypertension can predict future systemic hypertension.[31]

Left ventricular function

Patients with CoA may suffer from LV pressure overload, which can cause compensatory hypertrophy.[3] Ventricular hypertrophy can lead to myocardial fibrosis, associated with LV systolic and diastolic dysfunctions.[3] Histological studies have demonstrated predominantly subendocardial fibrosis of the LV, suggesting insufficient coronary flow (reserve) with compromised diastolic myocardial perfusion.[3] Although LV changes tend to normalise after CoA-repair, LV function remains subnormal in many patients.[3] Older age at the time of repair is associated LV hypertrophy and increased LV mass.[31 32 34] The ejection fraction and fractional shortening may be preserved in the early stages of ventricular dysfunction.[47] However, abnormal myocardial deformation indices (strain, strain rate and torsion) are found even in patients with well-repaired CoA, preceding systolic dysfunction.[3 44] Assessment of LV function is, therefore, important in CoA follow-up and should include myocardial velocity and deformation parameters.[3]

Pregnancy

Women with CoA have an increased risk of cardiovascular complications during pregnancy.[48] Hypertensive complications occur in 25% of pregnant patients with CoA.[48 49] Both pre-existing- and pregnancy-induced hypertension occur more often in patients with CoA, associated with adverse foetal outcome due to preterm delivery, low birthweight for gestational age

and admission to the neonatal intensive care unit.[49] Female patients with CoA with pregnancy wish should preferably undergo assessment of their haemodynamic status, severity of (re-)CoA and associated lesions before conception.[50]

CONCLUSION

Despite widespread availability of foetal cardiac screening in the current era, CoA often remains undiagnosed prenatally which poses significant risks of mortality and morbidity to the affected child. Prenatal screening for aortic arch abnormalities, adequate clinical examination and (if necessary) cardiovascular imaging of the neonate are, therefore, of great importance. Neonatal CoA is preferably repaired surgically, whereas BA and stent placement are the treatment of choice for older children and adults. Despite adequate treatment, patients with CoA have a reduced life expectancy and increased risk of cardiovascular complications later in life, related to hypertension, LV dysfunction and hypertrophy, restenosis, aneurysm formation and cardiovascular and cerebrovascular diseases. Timely intervention is crucial, as early treatment may prevent cardiovascular complications. Recoarctation should be proactively addressed. Cardiovascular complications may occur decades after initial treatment, warranting lifelong follow-up. Blood pressure controls should be performed yearly, and regular evaluation of the heart and aorta with echocardiography and/or CMR are recommended.

Contributors EJD collected data and wrote the manuscript. TL reviewed the manuscript and provided images. HBG served as scientific advisor and was a large contributor to the manuscript.

Competing interests None declared.

Provenance and peer review Not commissioned; externally peer reviewed.

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*Heart*2017 103: 1148-1155 originally published online April 4, 2017
doi: 10.1136/heartjnl-2017-311173

Notes