The long-term management of the patient with an aortic coarctation repair


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Introduction
Several areas of concern arise in the management of the adult patient who has had a surgical repair of an aortic coarctation. The aim is this article is to review these areas and make suggestions regarding current clinical practice.

Coarctation of the aorta accounts for 6.8% of congenital heart disease, with an incidence of one in 12,000 live births[1]. Coarctation is a heterogeneous lesion with variability in the degree and site of obstruction, the presence of collaterals, and associated cardiac defects including bicuspid aortic valve, patent ductus arteriosus, or ventricular septal defect. For this reason there is a marked diversity in the timing and nature of clinical presentation. Severe coarctation of the aorta most commonly presents in the neonatal period with cardiac murmurs, diminished femoral pulses and cardiac failure and intervention is often undertaken shortly after detection.

In the past, coarctation was considered to be a simple ‘correctable lesion’ and surgery, first performed in 1944[2], was believed to be curative. In keeping with this belief normotensive patients were often discharged post-operatively with no long-term follow-up.

Surgery
Since its introduction surgical correction has taken several forms. Patient age, morphology of the coarctation and transverse arch, concomitant cardiac lesions, and the personal preference of the individual surgeon have dictated the technique employed. Over the last 15 years, as increasing numbers of these patients reach maturity, additional information has become available on the long-term outcomes of the various procedures undertaken and this information now also influences the choice of initial surgery.

In infants and young children there are two main surgical techniques: subclavian flap repair and end-to-end anastomosis. Subclavian flap procedures utilize the subclavian artery to enlarge the aortic lumen and was, until recently, thought to be associated with a reduced incidence of restenosis when undertaken in early life[3]. End-to-end anastomosis, in which the aorta is extensively mobilized and the descending aorta is anastomozed to the underside of the arch, is especially useful in the setting of a hypoplastic arch. In larger children or adults this approach is often technically difficult owing to the undue tension created at the anastomotic site. In this situation Dacron tube interposition grafting or tube bypass grafting may be employed. Dacron elliptical patch grafts were performed in the 1970s and early 1980s. This procedure had the advantage of creating a large aortic lumen with a short cross-clamping period minimizing the risks of paraplegia. During the 1980s, however, it became apparent that this form of correction was associated with a significant risk of aneurysm formation[4]. Balloon angioplasty of native coarctation is a relatively new technique and in experienced hands is a valid alternative to surgical correction[5]. Its use in this role is not discussed further in this review.

Continuing care of the patient following repair
In previous years, if patients were clinically well and normotensive following surgery, they were often discharged from follow-up. This is now a cause of concern in the light of increasing evidence of the late complications associated with even the most effective repair.

Even if remaining under review, coarctation patients have been difficult to assess and until recently...
casual clinic blood pressure measurements, examination of the femoral pulses, and chest X-rays, have been the only methods of screening patients for potential problems. Unfortunately, significant changes in aortic morphology can occur with little in the way of examination findings.

Trans Thornton or transoesophageal echo image quality is often poor, with inaccuracies in Doppler estimations of repair site gradient. Difficulties arise in aligning the Doppler beam with flow and gradients are often underestimated, especially in patients with difficult anatomy. Cardiac catheterization, the gold standard, has been reserved for those in whom there is a clinical suspicion of a problem, but is too invasive to form part of routine review procedures.

Many of these problems have been superseded by magnetic resonance imaging. Magnetic resonance imaging is an excellent modality for demonstrating aorta morphology and has revolutionized the screening of coarctation patients for aneurysm formation. ‘Angiography-quality' images can be obtained safely and non-invasively using spin echo sequences. The magnetic resonance aortic ratio, that is the ratio of the diameter of the aorta at the repair site vs the diameter at the diaphragm, have been used to radiologically define those at risk of aneurysm formation or re-coarctation with a ratio of >1.5 or <0.9, respectively. It has been suggested that the finding of an aortic ratio >1.5 identifies patients at high risk of progressive anaerysmal dilatation over 3–5 years.

With regards to the diagnosis and accurate assessment of re-stenosis, cardiac catheterization is still more favoured than magnetic resonance assessment, especially if intervention is planned. Magnetic resonance does, however, fulfil the role of being a useful surrogate for catheterization when screening for re-stenosis and as techniques of gradient measurement improve this remit may be widened.

Decreased upper body vascular compliance is thought to be an aetiological factor in the development of late hypertension and magnetic resonance techniques have been used as a research tool to calculate aortic compliance in the proximal aorta. If initial observations are confirmed, future risk stratification of the post-correction patient may involve routine magnetic resonance assessment of compliance.

With the advent of widely available magnetic resonance imaging, a wealth of information regarding the ‘natural history' of these lesions is becoming available. During this learning phase, it could be argued that there is justification for 12–24 monthly magnetic resonance screening of all patients. In situations with scarce resources, priority must be given to patients with patch repairs or those with a clinical suspicion of either re-stenosis, dissection or aneurysm formation.

**Potential long-term complications**

As an increasing number of patients with coarctation repairs are now reaching adulthood, several specific areas of concern are becoming apparent.

**Post-correction hypertension**

Thirty percent of patients with surgically treated aortic coarctation become or remain hypertensive despite correction of their lesion with an increased risk of end-organ damage and accelerated atherosclerosis. Age at the time of repair is an important risk factor for the development of hypertension with early surgery being associated with an increased incidence of late hypertension. The development of late hypertension, that is occurring many years after correction, and exercise-induced systolic hypertension may go unrecognised especially if patients are no longer under regular review. The pathophysiology of these abnormal blood pressure responses is not fully understood. Neuro-endocrine parameters, including renin and catecholamine activation, functional re-coarctation, baroreceptor alteration and decreased aortic compliance have all been implicated. Hypertension once identified is effectively treated with either beta-blockers or angiotensin converting enzyme inhibitors specifically targeting the underlying neuro-endocrine abnormalities.

Measurements of clinic blood pressure every 6–12 months are adopted by most physicians as adequate screening for late hypertension, but more detailed blood pressure characterization with exercise testing and 24 h monitoring may be justified. Twenty-four hour ambulatory blood pressure levels may correlate more closely with end-organ damage than clinic blood pressures in patients with essential hypertension and subtle abnormalities in 24 h recordings have been documented in normotensive post-correction coarctation patients. There is no evidence at present to suggest that treating these abnormalities affects outcome either in terms of mortality or measurable intermediate endpoints, such as left ventricular hypertrophy. There would be justification in performing a multi-centred placebo-controlled trial of anti-hypertensives in this population, attempting to modify both 24 h blood pressure and left ventricular hypertrophy. Until clearer evidence is available, patients need to be assessed on an individual basis e.g. the threshold for commencing therapy in a patient with normal clinic blood pressures but an abnormal 24 h recording may be influenced by the extent of left ventricular hypertrophy, and aortic morphology.

**Bicuspid aortic valve disease**

Up to 80% of those with coarctation have bicuspid aortic valves. The majority of these lesions show pathological progression, although the changes may not reach the point of requiring intervention. Both aortic stenosis and aortic regurgitation can occur in this setting. Ascending aortic aortopathy with an increase in aortic root dimensions, aneurysm formation and dissection has also been documented. These features have led to the phrase ‘bicuspid aortic valve syndrome' to convey the
heterogeneous sequelae that may be associated. Consideration of aortic valve pathology is important when reviewing the coarctation patient and can be simply assessed both clinically and with echocardiography.

Re-coarctation

Re-coarctation or re-stenosis has long been identified as a post-correction problem. Invasive gradients of greater than 20 mmHg have been used both to define re-stenosis and to determine a cut-off point for intervention[24]. Since the early 1980s, balloon angioplasty has been used, as an alternative to re-operation, in the treatment of re-coarctation[25–32]. Balloon angioplasty in this setting is occasionally associated with deaths (0.7%), ilio-femoral trauma[33] and aneurysm formation at the angioplasty site. Despite the recognised complications, in experienced centres balloon angioplasty is a safe and effective technique with morbidity and mortality results superior to that of repeat surgery. The most significant disadvantage of angioplasty is that approximately 25% of patients experience only a short-lived improvement in the degree of obstruction. Repeated procedures can, however, be performed safely with excellent follow-up gradients maintained at 2–6.5 years[34,35].

Stenting of aortic coarctation is practised in a few centres for both native and re-coarctation. It is hoped this will have the advantages of being associated with lower rates of both re-stenosis and aneurysm formation[36]. As these techniques improve and move into routine clinical practice specific guidelines will evolve for both the indications for, and techniques of, intervention.

Aneurysm formation

One of the most concerning clinical complications affecting this group is the predisposition to aneurysm formation. This is particularly true for those who had primary correction, or repeat surgery for re-coarctation, using Dacron or Goretex patch grafts. This problem was common enough, occurring in up to 20% of patients[37], to convince many groups that such procedures should, in the main, be abandoned. Aortic aneurysms, whether post-surgery or post-angioplasty, may be asymptomatic with the first indication of their presence being aortic rupture. Even for this reason alone all patients with previous patch repairs should be recalled and remain under life-long review with regular imaging.

Pregnancy

Pregnancy in those with coarctation repairs is potentially problematic. In normal pregnancy, changes occur in aortic diameter and the aortic root dimensions increase[38]. This may be an aetiological factor in the increased risk of aneurysm formation and rupture reported in pregnancy[39]. The haemodynamic effects of pregnancy with increased cardiac output and circulating blood volume[40] may also have a deleterious effect on the pregnant patient with a coarctation repair. The true prevalence of hypertension and pre-eclampsia during pregnancy and details of fetal outcome in these patients is unknown as numbers are small.

Ideally all female patients of child-bearing age should be advised regarding the potential risks of pregnancy and those considering embarking on a pregnancy in the near future should have a detailed risk assessment. Clinical and echocardiographic re-assessment of concomitant lesions, especially bicuspid aortic valve disease, is necessary. Cardiac medications must be re-assessed. Angiotensin converting enzyme inhibitors, in particular, should be avoided due to fetal skull abnormalities and renal tubular dysgenesis[41]. Beta-blockers, although associated with intra-uterine growth retardation[42] are preferred for control of hypertension. It would be advantageous to postpone pregnancy if blood pressure control is suboptimal.

Baseline magnetic resonance imaging is useful in risk stratification of aortic dilatation and aneurysm formation when performed in the pre-conception period. Patients with aortic diameters >45 mm should be advised to avoid pregnancy and followed closely. Caution, at least at this stage when the history of these lesions is unclear, should also be exercised when counselling those with aortic ratios of >1.5. Pregnancy should ideally be postponed unless serial imaging reveals no evidence of progression.

Inevitably patients will present during pregnancy with no opportunity for pre-conception counselling and assessment. Magnetic resonance imaging, if performed after the first trimester, is thought to be safe and again is helpful for assessing all patients but is particularly important in those with patch repairs. Those assessed at being at higher risk should be encouraged to have their obstetric care at a centre with experience in caring for patients with congenital heart disease. Specialist anaesthetists, obstetricians, cardiologists and midwives are all potentially required to supervise shared care. A time of delivery caesarean section should be reserved for obstetric, rather than cardiac indications. However, labour should be as short and pain-free as possible with a low threshold for the use of forceps and epidural anaesthesia. Antimicrobial prophylaxis at delivery should not be forgotten.

Conclusions

Coarctation of the aorta, despite its successful surgical correction, is a condition that requires life-long follow-up in a centre with expertise in this field and adequate invasive and non-invasive imaging facilities. With the advent of readily available magnetic resonance imaging, balloon angioplasty, stenting and improved
surgical techniques there is great opportunity for improvement in the long-term management of this condition. Unfortunately, the shortcomings of previous therapies are also being revealed. Outlined above are our own areas of concern and proposed follow-up protocols. An 'over-zealous' approach may be necessary until we have a clearer understanding of the 'natural' history, and optimum treatment, for the problems discussed. Although the overall numbers of patients in these subgroups is small, with time and discussion we will hopefully be able to move from the field of anecdote to a more evidence-based practice. This will surely mean an improvement in the standard of care provided for future patients.

References


