EDITORIAL

Stenting the mildly obstructive aortic arch: useful treatment or oculo-inflatory reflex?

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Heart 2006;92:1541–1543. doi: 10.1136/hrt.2006.093302

The relative risks and benefits of the physical relief of mild aortic arch obstruction by angioplasty and stenting remain unclear

Despite many years of surgical and more recently percutaneous therapy for coarctation of the aorta, patients continue to suffer from hypertension and its complications in the long term. Because of a paucity of robust data, the optimum mode of treatment for patients with mild aortic obstruction and arterial hypertension remains unclear.

In this issue of Heart, Boshoff *et al* describe their experience of stenting hypoplastic aortic arch segments in patients with mild pressure gradients and arterial hypertension. The intervention was technically successful in all patients with the only reported complications being two moderate groin haematomas. There was an immediate reduction in invasively measured peak pressure gradient across the stented hypoplastic area, but many of the treated patients remain hypertensive at short term follow-up.¹

BALLOON ANGIOPLASTY AND STENTING

As equipment used for stenting aortic arch obstruction has become more refined and operators more experienced, balloon angioplasty and stenting for aortic coarctation has become commonplace, and has overtaken surgical intervention as the treatment of choice for aortic coarctation or re-coarctation, particularly in adolescents and adults in many centres. This change in practice is reflected by the profusion of case reports and relatively small series reports in the last 10 years.^{2–4} Employment of the technique is also supported by current National Institute for Health and Clinical Excellence (NICE) guidelines published in 2004.

Patients born with coarctation of the aorta die, and suffer more cardiovascular morbidity, at a younger age than their contemporaries, mostly due to the complications of hypertension. Disappointingly, even following apparently successful coarctation repair, arterial hypertension occurs in 20% of children and at least 25% of adults.^{5,6} Although young patients with hypertension have a low absolute risk of developing cardiovascular disease, they have a high risk relative to their contemporaries. Those in the top quartile of blood pressure aged 30 years can expect a significant rise in their blood pressure over the next 10 years, and tend to remain in the top quartile as they age. The current British

Hypertension Society guidelines recognise this and state that “although the hypertension in younger people may appear too mild to treat, it is not benign...it cannot be assumed that subtle and progressive vascular damage occurring in the younger hypertensive patient is necessarily reversible”.⁷

It is known that the adverse effect of hypertension on vascular complications can be modified by actively lowering the blood pressure. In patients with systolic blood pressure of 140–159 mm Hg, achieving a sustained 12 mm Hg decrease in blood pressure for 10 years will prevent one death for every 11 patients treated. Thus, while there is a paucity of data to guide management of hypertension in patients in their 20s and 30s (and it is unlikely that this will improve; as a group, these patients have such a low overall cardiovascular risk that they will contribute few end points to any outcome trials), extrapolation of the available data suggests that (age and gender adjusted) normalisation of blood pressure in these patients, by whatever method, should reduce their excess cardiovascular morbidity and mortality.⁸

So, how best to treat the hypertensive patient with mild aortic arch obstruction, usually in the face of a previously treated coarctation?

WHAT IS A “NORMAL” BLOOD PRESSURE IN A YOUNG ADULT?

Firstly, one needs to be able to identify the patient as hypertensive. There are a number of relatively large population based studies examining clinic and 24 hour ambulatory blood pressure profiles in a normal young adult population. Mean blood pressure, as assessed by 24 hour ambulatory monitoring, in women aged 20–29 years is as low as 108/68 mm Hg, and in men of the same age is 117/70 mm Hg.⁹ It should be recognised that a young patient whose blood pressure is well outside the 95th centile when adjusted for age can be deemed to be “normotensive” by current guidelines, a feature which may falsely reassure both patient and unwary clinician.

MEDICAL TREATMENT?

The mechanism of the development of hypertension in this group of patients is poorly understood, and most likely to be multifactorial, making tailoring pharmacological treatment difficult. Most clinicians treating these patients tend to select well tried antihypertensive drugs with very good side effect profiles, mindful of the facts that the patient may be on treatment for very many years, and that this otherwise fit and

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Published Online First
3 July 2006

active person will not be tolerant of adverse side effects. Anecdotally, hypertension in patients with coarctation (even after relief of physical obstruction) is often difficult to control despite large doses of a number of different antihypertensive medications.

PHYSICAL RELIEF OF RESIDUAL OBSTRUCTION?

It is clear from the current literature that if there is a significant degree of aortic obstruction, relief of this by whatever means reduces the blood pressure in the proximal segment. It appears that the reduction in upper limb systolic blood pressure gained by relieving the obstruction is slightly less than the reduction in peak to peak pressure drop measured invasively. For example, Eicken *et al* reported a median improvement in peak to peak pressure drop of 21 mm Hg following aortic stenting in 43 patients. This translated to a median improvement in systolic blood pressure of 16 mm Hg at follow up.¹⁰ Harrison *et al* reported a reduction in mean peak to peak pressure gradient of 42 mm Hg following stenting in 27 patients. This resulted in a reduction in mean systolic blood pressure of 34 mm Hg.¹¹

However, the blood pressure does not return entirely to normal in a large proportion of patients, particularly those who are older at the time of repair, demonstrating that merely relieving the physical obstruction alone does not necessarily "cure" the associated hypertension even when the physical result is technically excellent.

SURGERY OR CATH LAB?

The majority of patients with aortic arch obstruction are relatively young and fit, with arterial hypertension being their only "problem" and usually many years of good quality life ahead of them. It is important therefore that the benefit of any procedure they undergo can be clearly seen to outweigh the risks of having the procedure performed in the first place.

Surgical intervention for coarctation or re-coarctation has been performed for many years with great technical success. There are, however, clear downsides to this approach, particularly for young adult patients. A thoracotomy (particularly a re-do) is undoubtedly an unpleasant procedure to recover from. The surgery leaves a large and often painful wound, which may be accompanied by a significant pleural effusion. Surgery entails a trip to the intensive care unit, usually 5–14 days in hospital and three months convalescence. This three month period can put great strain on patients, their families and their employers. A percutaneous approach avoids a thoracotomy, involves only an overnight stay in hospital, and the patient should require less than a week off work.

Clearly, the main aim of any intervention is to "cure" the hypertension while experiencing the lowest possible morbidity and mortality risk. There has been only one randomised study comparing angioplasty with surgical repair for coarctation.¹² This study demonstrated similar complication rates, but a higher rate of re-coarctation and persistent hypertension were seen in the angioplasty group than in the surgical group. A recent meta-analysis has examined 16 published English language reports related to the treatment of adult and adolescent coarctation since 1995. While recognising the limitations of such meta-analyses, the author reports that primary stenting carried the lowest risk of complications, followed by surgery (odds ratio (OR) 1.3), followed by isolated angioplasty (OR 2.4). Reported restenosis rates after stenting were 0–25%, and after surgery were 0–9%. There was a higher risk of requiring repeat interventions after stenting compared with surgery (OR 16.1).¹³ It is clear from the available literature that the percutaneous approach is associated with more aneurysm formation at the site of

repair than surgical intervention. As many as 20% of patients undergoing coarctation angioplasty will develop aneurysms at the site. These are not necessarily large and in many cases require nothing other than a watchful eye. It seems likely that given the increased use of covered stents in this situation, this rather high rate of aneurysm formation will fall.¹⁴

Another issue which is often not considered is the radiation exposure experienced by these patients. Although the radiation associated with the percutaneous intervention is a "one-off", the dose from this procedure is often higher than that received during routine coronary angiography. It has been estimated that the radiation dose associated with coronary angiography causes 280 cases of cancer per million examinations. In addition, patients with stents will require long term follow up by computed tomographic (CT) scanning, because the artefact produced by the stent on magnetic resonance imaging makes assessment of residual obstruction and aneurysm formation impossible, and the radiation associated with a CT scan causes 60 cases of cancer per million examinations.¹⁵ This equates to around one cancer induced per 1000 patients treated, assuming a dose equivalent to coronary angiography for the procedure itself and 10 follow-up CT scans throughout a lifetime.

CONCLUSION

Unfortunately, despite patients undergoing invasive procedures to tackle coarctation of the aorta for many years, there is a paucity of outcome data to guide the long term management of hypertension in these patients. In the absence of long term outcome data, treatment decisions are currently based upon extrapolated data, clinicians' own experience and common sense on the part of both patient and clinician.

Physical relief of even mild to moderate degrees of obstruction of the aortic arch in patients who are hypertensive is likely to improve blood pressure control, although it must be recognised that many patients will remain hypertensive for their age and sex and will require additional medical treatment. As mentioned above, if one can achieve a sustained fall of 12 mm Hg in systolic blood pressure for 10 years, this can be estimated to prevent one cardiovascular death for every 11 patients treated. This beneficial effect must be balanced with the risks of the procedure itself, the lifetime risks of the radiation exposure required for adequate post procedural follow up, and the risks of re-intervention should it be required.

While a percutaneous approach seems just as efficacious in relieving obstruction and lowering blood pressure as surgery, there is undoubtedly a higher risk of aneurysm formation at the site of the intervention. Many adolescent and young adult patients will prefer to accept this risk and opt for a percutaneous approach rather than a surgical one because of the short length of stay and short recovery time.

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IMAGES IN CARDIOLOGY

doi: 10.1136/hrt.2006.092924

Coronary artery giant aneurysm with fistulas to the left ventricular diverticulum

A 71-year-old man was admitted for investigation of an abnormal cardiac silhouette on chest x ray film (panel A). He had no chest symptoms. Multislice computed tomography revealed a giant aneurysm formation (71 × 88 mm) slightly enhanced by contrast medium. The circumflex coronary artery ran over the surface of this aneurysm, but the communication could not be well defined (panel B). Selected left coronary angiography showed that the giant aneurysm received blood flow from the elongated circumflex artery (panel C). Left ventriculography showed that there was a diverticulum of the basal posterolateral left ventricular (LV) wall and that it was connected to the aneurysm via a vessel reversely contrasted in systolic phase (panel D).

Based on these findings, we diagnosed a coronary fistula originating from the circumflex artery associated a giant aneurysm draining into the LV diverticulum.

The patient underwent resection of the aneurysm. As expected, there were two holes connecting the circumflex artery and the LV diverticulum (panel E). Ligation of the distal circumflex artery and closure of the LV diverticulum with a Dacron patch were performed. The postoperative course was uncomplicated, and the patient has remained asymptomatic.

Coronary artery fistulas with a giant aneurysm are very rare, and, in most cases, drain to the coronary sinus, pulmonary artery or right ventricle. This is the first case report of a coronary artery giant aneurysm fistulating into the LV diverticulum.

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