The Adult with Repaired Coarctation of the Aorta

Eric M. Horlick, MDCM, FRCPC, Peter R. McLaughlin, MD, FRCPC, and Lee N. Benson, MD, FRCPC

Corresponding author
Eric M. Horlick, MDCM, FRCPC
University Health Network, Room 6E-249, 200 Elizabeth Street, Toronto, Ontario M5G 2C4, Canada.
E-mail: eric.horlick@uhn.on.ca

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Coarctation of the aorta is a common congenital lesion that may often be repaired or intervened upon early in life. The management of patients with this disorder revolves around the concept that although the coarctation may be treated, what remains is a diffuse systemic cardiovascular disorder. Careful clinical care and investigation is required to reduce morbidity from recurrent disease and residual lesions. The natural and modified history of the disorder is reviewed. This article focuses on the clinical care of adults with repaired coarctation and includes a review of clinical goals and investigation as well as indications for reintervention.

Introduction

Coarctation of the aorta is a congenital cardiac anomaly consisting of narrowing of the upper descending thoracic aorta which is sufficiently severe to cause a pressure gradient. The condition was first described in 1760 by Meckel and Morgagni [1,2]. The first coarctation repair in a patient was performed by Crafoord in 1944 [3]. The technique of subclavian turndown and end-to-end anastomosis described in 1944 and 1945 led to modifications including prosthetic patch grafts, subclavian patch aortoplasty, and prosthetic tube grafts [4,5]. For many years surgery remained the only alternative for therapy, until the first balloon angioplasty for coarctation of the aorta was reported in 1982 by Singer et al. [6]. Numerous reports of efficacy and outcomes of this procedure followed. Reports of the use of balloon-expandable stent placement to treat coarctation and re-coarctation demonstrated the potential of this method of treatment as a surgical alternative [7].

Adult cardiologists in specialized congenital heart disease (CHD) clinics, as well as those practicing in other settings, are now faced with the task of following both surgically and interventionally treated patients. The myth of the "complete repair" following a reparative intervention, as well as the lack of structure in the United States to transition patients from the pediatric world to adult cardiology, continue to be perpetuate a care gap that affects these patients. Patients are often unaware that follow-up is required and providers who see few of these patients are unsure how to advise them. The follow-up of these patients is often sporadic as they may be asymptomatic but harboring significant anatomic abnormalities. This article presents important information for the clinician caring for adult patients with repaired coarctation.

Incidence and Morphology

Coarctation is not an uncommon lesion; it accounts for 7% of patients presenting with cardiac lesions, occurs in one of 2323 live births, and ranks sixth in frequency of all congenital heart lesions [8–10]. Coarctations vary in severity and extent; 33% of autopsy specimens before repair show moderate luminal narrowing, 42% show severe stenosis, and 25% luminal atresia [11]. In addition to a diaphragm or shelf at the level of the arterial duct a variable degree of hypoplasia of the isthmus (the segment between the left subclavian artery and the insertion of the arterial duct or ligament) or transverse arch can be associated. At present the absence of a lumen is a strong argument for an initial surgical strategy, despite scattered reports of the interventional management of aortic atresia [12]. The presence of transverse arch hypoplasia is a variable finding and may similarly influence the initial decision to proceed with surgery. Occasionally an adult with evidence of previous unrelated surgical scars but who was lost to follow-up without an operative report may present with a redundant and severely kinked aorta opposite the ligamentum arteriosum without any pressure gradient, a finding called *pseudocoarctation* [13].

Histology

It is important to recognize that coarctation of the aorta is a diffuse process that cannot be simply resected or stented. Histologic study at the site of the diaphragm or

Table 1. Therapeutic approaches for coarctation of the aorta

Simple coarctation	Reoperation after previous repair
Prosthetic patch	Endovascular stent
Homograft	Balloon dilation
End-to-end anastomosis	
Tube interposition graft	
Endovascular stent	
Balloon dilation	

shelf has shown deformity and thickening of the media within the vessel wall [14]. In older children and adults, a thickened avascular intimal tissue overlies this media, composed of collagen layers, variable amounts of elastic tissue, and occasional smooth muscle cells [14]. Depletion and disarray of the medial elastic fibers at and around the coarctation site may be found, but are not a consistent histologic finding [15,16]. This appearance has been considered a potential morphologic substrate for weakening of the aortic wall, aneurysm formation, or dissection [17]. These findings are similar to the lesions of cystic medial necrosis described in idiopathic dilation of the aorta and in Marfan syndrome [15–17]. The aortic wall immediately distal to the coarctation is also thickened, and microscopy reveals localized intimal hyperplasia that can be seen with distortion of the medial wall architecture. The isthmus and transverse arch have the same histologic structure as the rest of the aorta. The diffusely abnormal arterial wall is a point of focus in the care of these patients. Surveillance for aneurysm formation both locally at the site of previous operative repair or intervention is relevant and the absence of a clinical gradient on examination should not constitute reassurance that all is well. Regular surveillance with cross sectional imaging is mandatory. The frequency of such imaging should be governed by the time elapsed since the last therapeutic procedure as well as the known presence of an already abnormal surgical or interventional site.

Associated Lesions

Several congenital abnormalities may coexist in about 50% of patients with the disease. The most common association is with a bicuspid aortic valve (22%–42%). The presence of a bicuspid valve is often associated with a diffuse aortopathy, which should be considered an additional potential risk factor when considering balloon angioplasty and stenting of coarctation in the adult [1,11,18]. The other associated lesions are more common in infants and children. The most common of these is a ventricular septal defect; however, a variety of other anomalies have been reported including abnormalities of the mitral valve apparatus, aneurysms of the distal aortic arch and intercostal vessels, a retroesophageal anomalous right subclavian artery, and intracranial aneurysms. Should endovascular therapy be required that may jeopardize the origin of a subclavian it is worthwhile to prove a patent contralateral verteberal artery during preprocedure testing [19]. In a number of series in the literature coverage of a subclavian with a stent graft or covered stent has often been asymptomatic but has occasionally presented as transient symptoms of arm ischemia or vertebrobasilar insufficiency that has infrequently required a later elective carotid to subclavian graft. An already compromised circulation to the arm from a previous operation would likely tolerate occlusion, given the prior development of collaterals. However, if the mammary artery has previously been used for coronary bypass grafting, or the contralateral subclavian is obstructed, collateral supply may be limited and preprocedure measures should be taken.

Natural History

Untreated aortic coarctation has a poor prognosis. Reifenstein et al. [11] reported 104 cases from the literature presenting in later life. The mean age at death was 31 years, with cardiac failure in 26%, at a mean age of 36 years. Aortic rupture (21%), bacterial endarteritis (18%), and intracranial hemorrhage (12%; 10 of 31 from a ruptured cerebral aneurysm) occurred at a mean age of 25 to 29 years. Older studies derived from postmortem findings are biased toward younger patients dying from complications derived directly related to the coarctation, whereas the deaths in older patients from other incidental causes or heart failure are underrepresented. Among all babies born alive with isolated coarctation who are untreated, 10% are expected to die in the first month from heart failure. A landmark study by Campbell [20] described a cohort of 181 patients followed for 716 patient-years in which 22 deaths were observed, corresponding to a mortality rate of 1.6% per annum for the first two decades, increasing to 6.7% per annum in the sixth and subsequent decades; 25% died before 20 years of age, 50% before 32 years, 75% before 46 years, and 92% before 60 years. Untreated patients surviving to adulthood typically had milder lesions and were likely asymptomatic for long periods, with arterial hypertension undiscovered until adolescence or adulthood.

Modified Natural History

Surgery

A variety of different operations were used to treat coarctation influenced largely by the age of the patient, the age at the time of repair, and the preference of the individual center. Table 1 summarizes a variety of strategies. The subclavian flap aortoplasty was used mostly in infants and children. The subclavian artery is split open longitudinally, transected, and after excision of the inti-

mal shelf, sutured across the coarctation site to enlarge the aorta. A second approach is resection and end-to-end anastomosis, which preserves the left subclavian artery, which is important in older children and adults. In some cases, a prosthetic patch aortoplasty was used to enlarge the aorta, and could be combined with resection and end-to-end anastomosis or subclavian flap aortoplasty. Occasionally when an end-to-end anastomosis is not possible, an interposition prosthetic tube graft is necessary, and occasionally an extra-anatomic correction is used with a conduit from the left ventricle or ascending to descending aorta.

Long-term outcomes after repair are quite consistent with other operations performed in CHD; they result in palliation without a cure, necessitating long-term and regular follow-up. The Mayo Clinic (Rochester, MN) experience was reported in 1989 with a median follow-up of 20 years [21]. Of 571 patients with long-term follow-up, 11% required subsequent surgery and 25% developed hypertension. There were 87 deaths at a mean age of 37 years. The most common cause of death was coronary artery disease followed by sudden death, heart failure, cerebrovascular accident, and ruptured aortic aneurysm. The age at initial repair was a critical predictor both of survival and development of hypertension. The series of Toro-Salazar et al. [22], which reported the 50-year postoperative follow-up of patients, had similar findings; 18% of those surviving surgery had died at a mean age of 34 years with 81% of patients alive at follow-up. Seventeen of the 45 late deaths were accounted for by coronary artery disease or a reoperation. In the Johns Hopkins (Baltimore, MD) study of survival up to 25 years after surgery, the mean annual mortality was 0.7% [23]. Older age at operation, particularly operation after age 40 years, is a significant risk factor for reduced late survival [23–25]. The modes of death included complications related to persistent or recurrent hypertension, rupture of intracranial or other aneurysms, acute aortic dissection, acute myocardial infarction, and complications of aortic valve disease. Persisting or recurrent coarctation and aneurysm formation at the site of repair are other late sequelae that may require further intervention after surgery [26–29]. A more recent Danish cohort of 229 patients who survived more than 30 days after repair showed late survival rates of 95%, 91%, 83%, and 69% 10, 20, 30, and 40 years postoperatively, respectively [30]. Late deaths were due to cardiovascular causes in 63% of patients; 25% of patients were on antihypertensive therapy. There was only a 60% freedom from cardiovascular complications, excluding hypertension, 30 years postoperatively.

Although outcomes have been significantly improved with surgical intervention, the burden of residual disease in the form of hypertension, residual lesions especially aortic valve disease, acquired vascular disease, and recurrent or persistent obstruction is significant and requires ongoing clinical surveillance and treatment.

Catheter interventions

Catheter intervention for treatment of coarctation has become increasingly applied as an alternative to surgical treatment. Balloon angioplasty was introduced in 1982 [6,31,32]. From a series of 422 native and 548 recurrent coarctation patients who underwent balloon dilation, it was noted that older age, recurrent obstruction, and higher pressure gradients were markers of a suboptimal outcome with this form of percutaneous intervention [33]. However, a recent 10-year follow-up of a childhood cohort, contrasting surgery and balloon dilation showed no differences in resting arm blood pressure, resting coarctation gradient, exercise performance, MRI measurements of the aortic arch, or need for repeat interventions between treatment strategies [34]. There was a higher incidence of aneurysm formation at the site of the intervention (35% vs 0%) and a greater difference in blood pressure between the right and left legs with exercise in the dilation group, with only 50% of those with angioplasty remaining free of both aneurysm formation at the site of the intervention and repeat intervention compared with 87.5% of the surgically treated subjects.

In adults, initial success rates were high, with reduction in arm-to-leg pressure gradients to less than 20 mm Hg. Restenosis was a significant problem, occurring in approximately 20% of older patients [7]. Late aneurysm formation has also been described with an incidence as high as 43% in early reports, and as low as 5% in later reports [7]. In adults, increasing age was identified as a risk factor for a suboptimal outcome [33]. Balloon angioplasty was first restricted to postoperative recoarctation because the periadventitial surgical scar tissue was thought to be protective against extensive vessel damage; however, large dissections and fatal aortic ruptures were described. With the development of large endovascular stents, balloon angioplasty with stent implantation came into use in the early 1990s. This technique was demonstrated to offer significant improvement in late outcome as compared with balloon angioplasty in the fully grown patient [35–39]. Fewer than 1% of patients require redilation of the stented segment [40–42]. The major risk of aortic rupture during the procedure persists with stenting, as well as a low but persistent risk of late aneurysm [36]. The introduction of the Cheatham-platinum polytetrafluoroethylene (CP PTFE)-covered stent, has significantly increased the safety of the procedure in preventing a major bleed from a through and through dissection, or "bailing-out" from a rupture after use of a bare metal stent. This stent has also been effective in treating postoperative aneurysm formation at the site of coarctation repair in those with bare metal stent strut fracture, and in situations where redilation of a previously undersized bare metal stent is thought to represent a particular hazard.

In a recent review by Carr [43••], the results of endovascular therapy (stenting and dilation) were compared with surgical techniques to repair coarctation in the adult patient. This review was heavily biased toward surgical therapy. Still, although the immediate improvement in

Table 2. Long-term risk factors after repair of coarctation of the aorta

Complication	Prevalence, %	Proportion of deaths, %	Risk factor
Coronary artery disease	5–23	25–66	Duration of prerepair hypertension, postoperative hypertension
Hypertension	25–75		Age at repair, recurrent lesion, length of follow-up, severe aortic regurgitation
Cerebral vascular accident	3	0–12	Pre- and postoperative hypertension, pre-existing Berry aneurysm
Heart failure		9–35	Hypertension, aortic valve disease, coronary artery disease
Recoarctation	3.1–10.8		Repair in infancy, subclavian flap repair, balloon dilation
Aneurysm/rupture	5.4–20	5–35	Balloon dilation, patch angioplasty
Endocarditis			Congenitally abnormal aortic valve

hypertension and the morbidity were similar across all groups, surgical therapy was associated with a lower risk of restenosis and recurrence, whereas endovascular therapy had a higher incidence of restenosis and the need for repeat interventions. Although the short-term results for endovascular interventions are highly compelling, the long-term outcomes of this treatment algorithm will need to be assessed in the future (Table 1).

Clinical Follow-up

Coarctation should be considered a lifelong disorder. It has been suggested that all patients with coarctation should have periodic follow up with an adult CHD specialist. The focus of the clinical encounter is to 1) identify and document modifiable coronary artery disease risk factors to optimize long-term outcome, 2) to identify acquired lesions (eg, coronary artery disease) with special relevance to the patient with coarctation, 3) to identify residual cardiac defects that may require operative or interventional repair, and 4) assess the site of repair and the remainder of the aorta for abnormalities that might require further therapy. Table 2 outlines the long-term risks associated with coarctation repair.

Before a patient is evaluated in our clinic we request previous operative or interventional repair reports and previous clinical records. The clinical history should identify and document risk factors for acquired vascular disease with special emphasis on hypertension and prior pharmacologic treatment. Symptoms of cardiovascular disease such as angina, congestive heart failure, syncope, and claudication are sought. Prior imaging studies should be reviewed and compared with recent studies whenever possible.

The physical examination focuses on the determination of blood pressure in all four extremities. The pathophysiology of persistent or late-onset hypertension, in the absence of persistent or recurrent coarctation or arch hypoplasia, is not well defined. Theories have included reduced aortic wall compliance, abnormal baroreceptor function, and endocrine activation. Subtle abnormalities can be detected in patients with normal resting blood pressures by ambulatory monitoring, and exercise testing [44,45]. Surgical and anatomic variants may influence the blood pressure measurements in the four extremities, so all should be measured and documented. For example, measuring blood pressure only in the right arm and leg in isolation may be misleading given the possible presence of an abnormal right retroesophageal subclavian artery. The left arm pressure may be reduced in the presence of a previous subclavian flap repair or if a stent has been placed over its origin. The right leg blood pressure may be reduced if a large bore access has been placed to deliver a covered stent or if a previous surgical cut down scar exists where a surgeon has gained access for femoral bypass. Thus, detection of a radial to femoral delay in the postoperative patient may not always reflect recurrent coarctation. Documentation of the presence or absence of pulses is important especially if an interventional strategy is planned.

Measurement of leg blood pressure is not standardized in patients with coarctation. Methods have included a manual mercury sphygmomanometer with auscultation over the popliteal artery, a cuff around the lower leg and palpation of the posterior tibial artery, the use of a cuff and Doppler of each site, or the use of an automated cuff. We favor the use of an automated cuff with multiple readings taken in each arm and leg and averaging the results. The auscultatory examination is critical to detect evidence of a congenitally abnormal aortic valve and residual ventricular septal defect. The presence of a murmur over the back with diastolic extension or the presence of a continuous murmur around the left scapula is unusual after repair.

The investigations relevant to patients with repaired coarctation may include some or all of those listed in Table 3. The electrocardiogram may demonstrate features of left ventricular hypertrophy and strain. A chest

Table 3. Diagnostic evaluation for the adult with repaired coarctation

Clinical examination
Electrocardiogram
Chest radiograph
Transthoracic echocardiogram
Exercise testing
MR or CT imaging of chest and brain
24-hour ambulatory blood pressure
Cardiac catheterization

radiograph may be the first sign of aneurysmal dilation at the site of previous intervention or of the aortic root. An echocardiogram is relevant to assess the aortic root dimension, the presence of a congenitally abnormal aortic valve and hemodynamically significant stenosis or regurgitation and to exclude other lesions, such as ventricular septal defect. Echocardiography is reasonably sensitive for the detection of recoarctation but notoriously poor for the detection of aneurysm of the repair site. Large adults are particularly difficult to image from the suprasternal window. The absence of significant collaterals in the postrepair group makes Doppler much more useful than in patients with native lesions [46]. Interestingly in a predominantly surgical population in the United Kingdom the most cost effective screening for complications in a cohort after surgical repair was a clinical visit and MRI [46]. However, MRI evaluation of the stented segment is particularly limited due to artifacts within the stent making them inaccessible for evaluation. In this population multislice gated CT angiography is a particularly useful imaging modality. We perform CT angiography on recently stented patients with hypotension, anemia, or an unusual amount of chest or back pain. A routine CT angiogram is performed 2 months postprocedure for all stented patients to evaluate the treated site and to rule out pseudoaneurysm formation.

We have been relying increasingly on ambulatory blood pressure monitoring to make clinical decisions in patients with treated coarctation. The benefits of this approach are reviewed in detail elsewhere [47•]. Two of the benefits include exclusion of white coat hypertension and determination of a daytime mean blood pressure. The use of exercise testing in adults after repair may be of special relevance in the detection of ischemia and documentation of functional capacity but is of questionable benefit for the assessment of blood pressure response. This is a controversial area. The interested reader is referred to a recent paper for a full discussion [48].

At 1 year after stenting, we repeat catheterization on all patients. The gradient across the stented segment is measured and angiography performed to exclude aneurysms and other zones of obstruction in the arch and isthmus. Stent integrity is assessed. Hemodynamics are measured without the limitations imposed by anesthesia at the time of the index procedure which tends to significantly lower systemic blood pressure. An angiographic assessment of the femoral artery previously used for stent delivery is performed. Coronary angiography is carried out in patients with markers of risk. Finally, all adult patients undergo CT angiography or MR angiography evaluation of the brain to exclude berry aneurysms.

In counseling patients we suggest lifelong endocarditis prophylaxis. Patients normotensive at rest and with exercise should lead normal lives. Those patients without recurrent arch obstruction, but resting hypertension, or with abnormal ambulatory blood pressure monitoring require medical management. Patients with recurrent anatomic obstruction and hypertension should be considered for reintervention.

Clinical Decision-making in the Adult Who Requires Reintervention

If the diagnosis of recoarctation is confirmed and the lesion characterized, the decision to intervene is dependent upon the severity of the lesion, the patient's physical status, and associated comorbidities. In our institution reintervention is almost entirely a catheter-based approach, with the exception of very large aneurysms or in the presence of anticipated technical difficulties, which may include significant arch hypoplasia with involvement of the great vessels not readily amenable to stenting or extreme aortic torutuosity. The decision to proceed should depend on a balance of procedural risk at a particular institution against the severity of the coarctation. With regard to recoarctation, the gradient at which to recommend intervention is required is not precisely defined. However, a gradient of 30 mm Hg or more is generally accepted by all as significant. In the pediatric literature, a gradient of more than 20 mm Hg is the threshold for intervention in the asymptomatic child, whereas some investigators would suggest that a gradient of more than 10 mm Hg in the adult, with a stiffer ventricle, is justification to intervene [49]. As a guide the following can be considered: all symptomatic patients with arm to leg gradients of more than 20 mm Hg should have some form of repair. The asymptomatic patient, with a 20-mm Hg gradient or more, with resting hypertension or left ventricular hypertrophy should be considered for repair as well. The procedure with either a bare metal stent or covered stent will eliminate the gradient, or reduce the gradient to a minimal and acceptable level in nearly all cases [33]. The risk is considered low and similar to surgical repair [43••], if one exercises careful patient selection. However, as with most interventional procedures there is some risk even with experienced operators. It is important to be cognizant of the risk factors for complications. These include the presence of a pre-existing aortopathy, particularly associated

with bicuspid aortic valve [50], the elderly patient [39], and recoarctation associated with synthetic patch repairs. In adults in whom concomitant coronary artery bypass graft surgery is required, an anterior midline approach is required, making access to the descending aorta somewhat more difficult. In these cases, an interventional approach to the coarctation may be chosen prior to the bypass graft surgery as a means of simplifying the surgical approach. We have treated those patients with aneurysmal dilatation at the site of previous surgery or stenting with coil occlusion and most recently with covered stents with success. The threshold for intervening on an aneurysmal segment relates to the absolute dimension as well as its progression over serial studies.

Pregnancy

Counseling prior to pregnancy should be advocated in patients with coarctation. It allows for the opportunity to identify those patients who might benefit from an intervention prior to becoming pregnant and to provide counseling to the parents on the risk of recurrence in the offspring.

Pregnancy is generally well tolerated. Several recent series have been reported. There are no data to suggest an increased rate of infertility in these patients [51]. There may be an increased risk of spontaneous abortion with reported rates of between 9% to 18% versus a 10% rate in normal subjects [52]. Sustained hypertension does not appear to play a role but the high recurrence rate in the offspring of patients with left-sided obstructive lesions may contribute (ie, left-sided lesions in the offspring are related to increased rates of pregnancy loss). Hypertension is more common in women with repaired coarctation (18%–30%) when compared with the general population (8%) as is preeclampsia (26% vs 3%) [52]. It is interesting to note that in the series from the Mayo Clinic there was no difference in the rate of miscarriage or Caesarian section, median infant weight, or hypertensive disorder in those who were repaired versus those with native coarctation. Aortic dissection was rare in the largest reported series but has been reported in the literature [53]. The low incidence is reassuring given the changes in the aortic wall (cystic medial necrosis) which are similar to those of Marfan syndrome. However, women with Turner's syndrome who become pregnant through in vitro fertilization may be at higher risk. Careful monitoring of blood pressure throughout pregnancy is mandatory and although evidence is lacking, the use of a β-blocker seems a reasonable choice of therapy in this population. There was a very high Caesarian section rate in the Mayo Clinic series, which is suspected to be on the basis of perceived obstetric risk. A vaginal delivery is suggested for most patients with CHD, preferably with epidural anesthesia. An assisted second stage to avoid maternal pushing or conversion to operative delivery may be required in a minority of patients with uncontrolled hypertension, aortic root dilation, or severe recoarctation. Antibiotic prophylaxis should be administered as per standard obstetrical practice.

Berry Aneurysms

There has been a long-known association of coarctation of the aorta and berry aneurysms of the Circle of Willis. This association also exists for those patients with Marfan syndrome and Ehlers-Danlos syndrome type IV. The association was first noted by Epinger in 1871. Forty years later Dr. Maude Abbott reported on 200 cases of adult coarctation in the presurgical era, noting that after congestive heart failure and aortic rupture neurologic compromise was the most common cause of death [54]; 10% of patients in her series had intracranial hemorrhage and 25% of these were cause by intracranial aneurysms.

A series of 277 patients with the diagnosis of coarctation seen at the Mayo Clinic were contacted and were invited to have a cranial magnetic resonance angiogram to identify the presence or absence of saccular aneurysms [55]. One hundred patients had scans revealing a 10% incidence of aneurysms with a mean diameter of 3.5 mm (range, 2–8 mm). Nine of the 10 patients were asymptomatic; one had had a previous intracranial hemorrhage managed by arterial ligation. One patient who had an aneurysm of the basilar artery underwent surgical ligation. The rate of aneurysms in the general population is between 1% and 5%, with the majority of these being small; the risk of subarachnoid hemorrhage is one case per 10,000 patients in the general population.

In a retrospective study examining 2621 patients with unruptured aneurysms without CHD it has been found that the rate of rupture is 0.05% per year [56]. The rate was much higher in those with previous bleeding and in those with aneurysms greater than 10 mm, and those affecting the basilar apex or the posterior communicating artery. In the prospective part of the same study 1692 patients who had aneurysm of less than 7 mm had a 0% 5-year rupture rate. The tailoring of therapy for an individual patient is complex and the choice of whether surgical clipping or endovascular therapy should be performed is likely best left to a specialized neurosciences group likely to involve one or more neurosurgeons, neuroradiologists, and neurologists. The benefit of routine screening for cerebral aneurysms and prophylactic treatment in patients with a history of coarctation remains to be proven but is of definite interest.

Conclusions

Coarctation of the aorta is a systemic disease that requires lifelong follow-up by experienced providers aware of the potential problems and issues encountered by these patients. Follow-up by specialists in adult CHD is optimal, as is a multidisciplinary team to deal with complex care issues. The

false notion that a prior surgical repair is always curative can be harmful in that the patients may not seek follow-up care. With expert care and attention to ongoing issues, these patients may enjoy the benefits of longevity and an excellent quality of life. Clinical encounters should focus on identification of relevant untreated or recurrent abnormalities and appropriate evaluation and treatment of risk factors for acquired disease. The possibility of requiring further interventional or surgical therapy requires discussion and referral to those with experience in caring for this complex disorder.

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