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Abstract

Coarctation of the aorta includes a wide array of anatomic and pathophysiologic variations, which may cause important long-term morbidity and mortality. Percutaneous techniques such as balloon dilation and stenting allow safe decrease or abolition of most of the gradients along the aorta; however, there are some limitations. Interventional techniques allow adequate stretch or therapeutic tear of the vessel wall while keeping complications such as an excessive tear, dissection, aneurysm formation, or rupture to a minimum. The interventional techniques are determined by characteristics of the patient such as age, size, and growth potential and the lesion such as degree of narrowing, length, and angulation and by local regulations and facilities.

Keywords

Aneurysm • Aortic arch • Balloon angioplasty • Covered stent • Hypertension • Rupture • Stent • Stent graft • Therapeutic tear

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Introduction

Diseases of the aorta include predominantly abnormal narrowing and hypoplasia, but also dilation and aneurysm formation. Coarctation of the aorta consists of a wide array of anatomic and pathophysiologic variations. It occurs with a frequency of 0.04 % of live births and comprises about 7 % of known congenital heart disease [1]. Narrowing of the aortic arch predisposes patients to high blood pressure and subsequent development of premature atherosclerosis, which may lead to myocardial infarction and stroke at a younger age. Long-term follow-up studies, even after successful coarctectomy, have shown significant morbidity and mortality beyond the fourth decade, usually secondary to or associated with arterial hypertension [2]. The aim of treatment should always aim for an aortic arch free of any gradient in order to avoid arterial hypertension and should improve long-term outcome in these patients. A blood pressure gradient at rest of greater than 20 mmHg between upper and lower extremities generally indicates the need for intervention. Additionally, pressure differences of less than 20 mmHg may warrant intervention if there is hypertension at rest and, especially, if there is left ventricular dysfunction or progressive hypertrophy. Lastly, exercise-induced hypertension is increasingly being recognized as another indication, as late outcome is also suboptimal [3].

Mechanism of Pressure Loss Along the Aorta

Several lesions, either isolated or in combination, may cause a gradient in the aorta, each with specific opportunities and limitations for transcatheter treatment:

- Discrete narrowing, typically a coarctation
- Tubular hypoplasia at:
 - The isthmus, distal to the left subclavian artery
 - The distal cross, between left carotid artery and left subclavian artery

- The proximal cross, between brachiocephalic trunk and left carotid artery
- The lower thoracic or abdominal coarctation (middle aortic syndrome)
- Steep angulation of the arch as seen in a cervical arch (between ascending aorta and proximal transverse segment)
- Any tortuous course of the arch
- An atretic segment, when acquired later in life, typically at the isthmus between the left subclavian artery and the coarctation site
- A postsurgical narrowing, which may consist of residual hypoplasia or stenosis, lack of (catch-up) growth of bordering segments, excessive scar tissue, a circular non-resorbable wire, or a tube interposition graft
- A residual, recurrent, or growth-induced narrowing after catheter intervention

Treatment of Coarctation

In 1944, Dr. Clarence Crafoord and Dr. Robert Gross performed the first successful surgical repair of coarctation of the aorta [4, 5]. During the ensuing four decades, surgery remained the only treatment for coarctation. In the late 1970s, percutaneous balloon angioplasty was described as an alternative to surgical repair [6]. Since then, transcatheter interventions have become increasingly popular and in many cases have emerged as the treatment of choice [7, 8].

Percutaneous Options and Mechanism of Action

Transcatheter options now include simple balloon angioplasty, angioplasty followed by stenting, primary stenting using bare or covered stents, and rescue stenting with a covered stent. The mechanism of relief involves stretching and tearing of the vessel wall.

By *simple balloon angioplasty*, the minimum a balloon will do is *stretch* the target lesion; however, this may be followed by early and late recoil of the lesion, making this technique less

satisfactory. A sustained result of balloon dilation typically involves a *tear* of the intima and partially the media while keeping the adventitia intact; such a therapeutic tear should be contained to the narrowed zone [9–11]. In order to obtain a sustained result, some degree of overdilation with the balloon is frequently required. After dilation, some mechanisms will limit or improve the long-term result: recoil and retraction of nongrowing scar tissue will result in recurrence of the narrowing or stenosis, but catch-up growth and remodeling may occur by release of the stenotic ring and enhanced antegrade flow. However, such tears may predispose to possible complications. These include dissection, false aneurysm, and rupture [12]. A *dissection* is a tear that extends beyond the coarcted segment in the axial dimension, permitting contrast to track extraluminally in a proximal or distal direction. The false lumen of such dissection may be progressive and cause distal tearing of the vessel wall or even occlusion of side vessels. A *false aneurysm* may result from a defect in the aortic wall, with contrast extravasation beyond the adventitial plane with a discrete length; such a false aneurysm may “grow” over time and eventually rupture. An *aortic rupture* is a frank disruption of the aortic wall, which appears angiographically as extravasation of contrast beyond the confines of the aorta into the mediastinum or pleural space. A relatively high incidence of aneurysm formation of between 2 % and 20 % has been reported after balloon dilation [7, 13]. Several techniques have been described to improve the hemodynamic result and reducing the risk for these complications: these involve low-pressure balloon dilation and interrogation of the stenosed site, progressive and/or stepwise dilation with noncompliant balloons in one or more sessions, and limiting the size of the balloon based on the narrowing itself or adjacent segments.

Since 1989, *bare metal stents* have been used to treat narrowing in the aorta [14–18]. Stents will overcome many of the shortcomings of simple balloon dilation. A stent will expand and scaffold the target region, thereby avoiding recoil and residual or recurrent stenosis. A good result can

be obtained by simple stretching of the wall without a tear as there is no need for overdilation. Stenting may therefore result in lower vessel wall complications: fewer aneurysms, no dissections within the stent as they are automatically contained by sealing the intimal flap, and less rupture. Where intimal tears occur, the stent provides a surface for formation of neointima over the tear and reinforces the weakened areas within the aortic wall reducing the risk of a false aneurysm. However, stent implantation has some shortcomings: the technique is technically more demanding and requires a bigger sheath causing more vascular trauma at sheath insertion point, the foreign metal may induce interactions with surrounding tissues such as coagulation, wall hyperplasia, sharp edges of the stents may protrude and damage the vessel, the stents alter local vascular compliance and impede vessel growth, and stents may fracture and collapse [19]. Overall, the use of bare stents has improved results while lowering the complication rate of vessel wall trauma to 1–5 % [20].

Initially, stent implantation was used only for cases where surgery and balloon angioplasty had failed. However, as experience increased, stenting gradually became the treatment of choice in selected patients with aortic coarctation [21]. This is especially the case when coarctation coexists with hypoplasia of the isthmus or transverse arch, or when balloon dilation has a high failure rate such as in a tortuous coarctation, a long segment coarctation, or mild discrete coarctation. Stents are particularly helpful in some postsurgical patients [22] where a non-resorbable wire was used: balloon dilation alone may only tear the intima from the vessel with possible major dissection before breaking the wire. In adult patients, stenting is now considered as the treatment of choice in any variant of aortic coarctation. At the other end of the scale, in children less than 10 years of age, it is preferable to avoid stenting, as several redilations may be required until the child is fully grown.

The availability of *covered stents* has further reduced the incidence of aneurysm formation and vessel rupture to less than 1 % as the covering

will seal any tear in the vessel wall [23]. Covered stents can also exclude an unwanted passage to vessels such as an arterial duct or an existing aneurysm [24–26] and allow creating a new vessel segment as in aortic arch atresia [27]. Covered stents typically will require a larger sheath of more than 1 F than the size needed for bare stents, may cover origins of side vessels, may cause flow obstruction if incompletely opened or partially collapsed, and if they migrate to an unwanted location, may be more hazardous than bare stents. Currently, the discussion is still open as to whether covered stents should be used as a routine or only in case of a complication or specific indication. In countries where covered stents are available, there is a clear shift towards more routine use of covered stents in order to reduce the early and late complication rates [28]. Having covered stents available in the catheter laboratory adds to the safety of any aortic procedure: bleeding due to excessive vessel damage can usually be controlled with a covered stent as bail-out procedure.

Equipment

Balloons for Angioplasty

Many balloons are currently available for angioplasty of the aorta. The differences between balloons include availability in different sizes and lengths; tapering of balloons (the shorter the better, as the nose of the balloon may cause unwanted lesions in the arch); compliance; profile which determines sheath size for vessel entry; mode of refolding, which determines sheath size for removal of the balloon and the extent of damage at distal point of sheath; nominal pressure; burst pressure; inflation and deflation time; resistance to puncturing (stent or calcium); mode of rupture such as a point, longitudinal, and transverse; coating of balloon and shaft properties, and stretchability on retrieval. It is not uncommon to have different types of balloons available on the shelf at all times.

Balloons for Stent Delivery

Many balloons are available for stenting of the aorta. Although the same balloon as the ones used for angioplasty may be used, each technique will determine different ideal requirements of the balloon. For stent delivery, additional features such as the balloon size, material, surface, and mode of inflation are important. A shorter balloon will avoid excessive shoulders on inflation which may cause flaring of stent and puncture of the balloon and will enhance stability of the balloon during inflation with reduced inflation time, and less hemodynamic effects, but may make slipping of the stent off the balloon more likely. The balloon material should be non-slippery and puncture resistant, especially when used with stents with “sharp” edges of the crimped stent; sufficient lumen should remain to allow symmetric inflation of the balloon as asymmetric inflation may result in “milking-off” the stent from the balloon during expansion. Several balloons are currently available from different manufacturers: Powerflex[®] and Opta Pro[®] (Cordis, USA), Z-Med[®] and BIB[®] (NuMED, USA), Cristal[®] (Balt, France), and many others.

In the early days, all stents were hand-crimped on single large diameter balloons for expansion and delivery within the coarctation. Large diameter single-balloon catheters tend to expand first at their ends and thereby evert the stent ends such that they protrude radially into the vessel wall, which predisposes to aneurysm or dissection at the edges of the stent. An important development of equipment for delivery of large diameter stents has been the Balloon-in-Balloon (BIB[®]) catheter (NuMED, USA). These catheters have a small inner balloon and a 1 cm longer outer balloon that is twice the diameter of the inner balloon. The BIB catheters offer the important advantage of opening the stent more uniformly along its length. They do, however, require a larger arterial sheath for introduction: the profile of BIB catheters with outer balloon diameters of 8–14 mm is 9 Fr, of 16 mm is 10 Fr, of 18–20 mm is 11 Fr, and of 24 mm is 12 Fr sheath. Thus, while BIB catheters prevent stent flaring, cause less stent

Table 64.1 Characteristics of currently available stents: crimpability, distensibility, foreshortening

	Min inner diameter (Fr)	Stent thickness (Fr)	Min outer profile (Fr)	Balloon premounted	Stent maximal diameter	% shortening at 20 mm
<i>Bare stent</i>						
Genesis XD	5	1	6		18.5	57
Valeo	P		6	≤10	20	50
AndraStent XL	8	1	9		25	25
AndraStent XXL	9	1	10		32	5
IntraStent LD Max	9	1	11		26	3
Cheatham-Platinum	7	2	9		28	18
Mounted CP	P	2	10 12	12–24	28	18
<i>Covered stent</i>						
CCP	7	2–3	10		26	18
Covered Mounted CP	P		12 14	12–24	26	18
V12 Advanta	P		10	≤16	20(22)	17

Legend: P = premounted; V12 Advanta can be dilated up to 22 mm, but recoils to <20 mm

foreshortening, allow repositioning after inflation of the inner balloon, and offer more precise control over stent placement without danger of “milking-off” the stent during inflation, single-balloon catheters are preferable in smaller patients to reduce the risk of injury to the femoral artery at the access site. In order to keep the sheath size as small as possible in smaller children, a single low-profile balloon may be used to deliver the stent and anchor it across the lesion and then further dilate the stent with another noncompliant high-pressure balloon.

Stents

Many stents are available for stenting the aorta. Differences between the stents include crimpability with low profile, distensibility, the pressure required to open and deploy the stent, conformability over the full length, foreshortening and radial strength at different diameters, flaring at the ends, open or closed cell design which determines longitudinal grip and side branch accessibility, sharpness of the edges and the wire within the stent, collapse resistance, wire fracture resistance by metal fatigue or intentional by balloon dilation, membrane covering, radiolucency, and MR

compatibility. Less important stent features for aortic application are flexibility and cell area which determines tissue prolapse through the cells. The following balloon dilatable stents are available to treat congenital lesions in different parts of the world: Palmaz[®] XL 10-series and Genesis[®] XD (Johnson & Johnson, USA), AndraStent[®] XL and XXL (Andramed, Germany), IntraStent[®] LD Mega and Max (ev3, USA), and the Cheatham-Platinum[®] (CP) stent (NuMED, USA). The Valeo[®] stent (Bard, USA) and Mounted CP stent[®] (NuMED, USA) are premounted, the latter within a sheath. Covered stents are available as the Covered CP[®] (CCP stent) or the Covered Mounted CP[®] stent (NuMED, USA), the V12 Advanta[®] stent (Atrium, USA), or can be handmade [29]. Some comparisons between stents are summarized in Table 64.1. Occasionally, self-expanding stents may be used, but these have a limited role in pediatric cardiology practice when treating narrowed segments. Such stent grafts typically require 22–24 Fr sheaths and are used to treat true aneurysms in adults.

Crimpability of the stent to a low profile determines the minimum sheath size, but this is inversely related to its distensibility, foreshortening, radial strength, and fracture resistance (Table 64.1). The Genesis and the

premounted Valeo stent have a low profile of 6 Fr when mounted on a balloon up to 10 mm, and can be dilated up to 18–22 mm, however, with significant foreshortening (>60 %) and fracture rate. Most other stents will require a minimal sheath size of 9–10 Fr, depending on the type of balloon used. The size of an adult aorta varies between 16 and 20 (and may be up to 22 mm), and so sheath size may be up to 14 Fr. In very premature infants, coronary stents can be introduced through a 4 Fr sheath and can eventually be dilated up to 5 mm and so currently these need subsequent surgical removal. The pressure required to open and deploy the stent and the sharpness of the stent wire will determine the required thickness and pressure resistance of the balloon, which determines the profile of the balloon and thus the final profile for insertion. Some conformability is desirable to allow the stent to conform to the curvature of the arch to reduce the likelihood of stent damaging the vessel wall. When partly covering the origin of a head and neck vessel, flaring the end of the stent into the side vessel may be desirable, especially when using a covered stent. Stents may have an open or closed cell design, which determines the maximal size of opening to a neck vessel.

A stent ending with a stiff thin circular wire will offer more grip, but this is not required in the aorta as the stent is usually well fixed by the stenosis, but the wire may cause more vessel damage, both at placement and at redilation, resulting in dissection or aneurysm formation at the ends of the stent.

Guidewires

Most operators use a long stiff 0.035" guidewire with a soft tip. The guidewire may be positioned in the ascending aorta or the left ventricle; but in order to avoid damage to the left ventricular apex, the aortic valve or a coronary artery, the tip of the guidewire should be curled. Positioning the guidewire in the right subclavian artery may offer more stability during stent deployment, but with some risk of dissection of the

brachiocephalic trunk [30]. The left subclavian artery is rarely used due to insufficient space for balloon inflation.

Sheaths

Hand-crimped balloon mounted stents should be delivered through a long sheath, keeping the stent covered until positioned across the stenosis. There are several appropriate types of sheath, the most popular being the straight Cook RB-Mullins design sheath (COOK, Denmark), which has a competent hemostatic valve, a radiopaque tip, and a side arm, allowing hand contrast injections during stent positioning. Some covering or a short, cutoff piece of sheath tubing placed over the stent is usually required to pass the stent safely through the valve.

Inflation: Speed by Hand or by Indeflator

Early in everyone's experience, it was felt that the balloon should be inflated as fast as possible to keep the hemodynamic burden on the heart as short as possible. However, this has often resulted in asymmetric expansion and migration of the stent during inflation. Most operators now recommend slow balloon inflation initially, allowing both shoulders of the balloon to rise around the stent, preventing the stent from being "milked" off the balloon. Once in satisfactory position, further balloon inflation may be much faster. The use of a BIB balloon has nearly abolished the risk for asymmetric stent opening and stent dislodgement.

The balloon can be inflated by hand, allowing the operator to continuously observe the stent and the coarctation site, and thus control the dilation, when opening the stent inside the lesion and stretching and tearing the stenotic region. The operator should know by experience the level of pressure that can be achieved with different syringes, keeping in mind that the larger the syringe size, the lower the maximal pressure that can be

achieved by manual inflation. For example, a 10 cc syringe typically will allow 6–10 atm pressure to be achieved while a 20 cc syringe allows pressure of 4–6 atm. An indeflator device may help achieve better control of the pressure, but ideally one operator should focus on the manometer while another operator focuses on the stent expansion. Most lesions will open with inflation pressures below 6 atm, but when higher pressures are required, an indeflator is necessary.

Stability of Stent

Adequate stability of the stent on positioning depends on the delivery ensemble such as stiff guidewire, stiff balloon shaft, and long stiff introducer sheath placed just below the balloon. Guidewire position may influence stability such as ascending aorta versus subclavian artery position of the guidewire. Often rapid right ventricular pacing at a rate of 180–240 bpm is used [31]. Rapid pacing may be of importance in adults with a high stroke volume and in particular in the presence of aortic regurgitation, or when deploying a stent in the transverse aortic arch closer to the heart. In exceptional cases, additional stability can be achieved by snaring and externalizing the distal end of the guidewire from the right radial or brachial artery to form a stable circuit for stent deployment [32].

Technique of Balloon Angioplasty

The procedure is performed under general anesthesia or heavy sedation as stretching or tearing the aorta is extremely painful for the patient. Vascular access is obtained via the femoral artery. An aortogram is performed with maximal elongation and profiling of the aortic arch. Initially, the plane of the aortic arch is determined. With the catheter in the ascending aorta, the frontal camera is placed in RAO projection until perfect alignment of the ascending and descending arch is obtained. Subsequently, the lateral camera is rotated in LAO projection at

exactly 90° to the RAO camera. From this angle, the aortogram will be perpendicular with maximal elongation of the arch. Angiography can be performed using a pigtail catheter with 1 cm radiopaque markers, allowing accurate calibration and exact measurements of the aorta. Alternatively, a Multi-Track® catheter (NuMED, USA) advanced over a 0.035" exchange guidewire can be used. This catheter has a 1 cm marker for accurate calibration and a monorail system allowing pullback gradients to be measured across the coarctation without losing guidewire position. The measurements of the aorta include systolic diameters of the distal transverse arch just proximal to the origin of the left subclavian artery, the aortic isthmus just distal to the origin of the left subclavian artery, the site of the coarctation, and the descending aorta above the diaphragm.

Different protocols have been reported to select the appropriate balloon size for safe dilation of the coarctation. Balloon size is limited by the neighboring segments such as the transverse aortic arch, the proximal isthmus, or the thoracic aorta at the diaphragm level and/or by the coarcted segment itself. The balloon should be no more than 300 % of the minimal diameter, provided this is well visualized by the aortogram. A low-pressure inflation is recommended to interrogate the compliance and narrowing of the vessel. If the stenosis was underestimated, a smaller balloon may be preferred for initial dilation. Progressive dilation can be performed, each time preceded by measurement of the residual gradient and a repeat aortogram to assess the result (Fig. 64.1). Not surprisingly, a bigger balloon will yield a better gradient relief, but may increase the complication rate.

For a patient younger than 1 year of age, balloon angioplasty is frequently only palliative as the rate of recoarctation has been reported to be more than 50 % [14]. For a patient older than 1 year of age, balloon angioplasty has reasonable immediate results, but the rates of recoarctation are still about 26 % [15].

Late false aneurysms have been reported in 1.5 % of cases after balloon dilation of a postsurgical coarctation [7], whereas they seem to occur

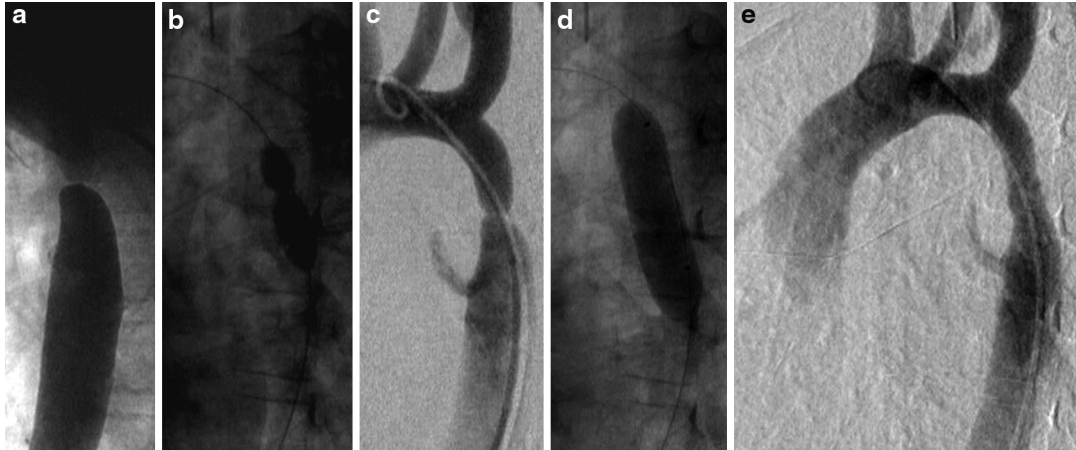


Fig. 64.1 Balloon dilation of native aortic coarctation. Six-year-old patient with a tight aortic coarctation: (a) Aortogram in the descending aorta shows narrow discrete coarctation. (b) Through a 5 F sheath, balloon dilation shown with an 8 mm balloon. (c) Repeat angiogram shows

residual discrete narrowing. (d) Further dilation with 10 mm balloon. (e) Final angiographic result with gradient reduction from 30–8 mmHg. At latest evaluation 2 years later, satisfactory growth of the distal arch without clinical gradient

more frequently (8–35 %) [13] in patients with a native coarctation. The lower incidence of aneurysms in postsurgical coarctation patients may be due to the fact that the aorta is surrounded by postoperative fibrosis and that most of the abnormal aortic wall tissue is removed at the time of surgery.

Technique of Stenting

The procedure is similar to balloon dilation in many aspects (Fig. 64.2). In the presence of a very tight or nearly atretic aortic segment, access may be needed via the right or left radial or brachial artery, allowing a catheter or guidewire to be passed from above through the coarctation site into the descending aorta. Rarely, transcatheter or a carotid arterial access via a surgical arteriotomy may be required. Heparin at a dose of 100 IU/kg is given after access is obtained; the activated clotting time is maintained above 220–250 s throughout the procedure, particularly due to the risk of long clots developing in the long introducer sheath. A Perclose (Abbott Vascular) suture can be inserted at this stage for rapid and lasting hemostasis at the end of the procedure. However, in

adolescents and young adults, good hemostasis can be achieved by prolonged manual compression under anesthesia even after removing a 14 Fr sheath. A 5 or 6 Fr end-hole catheter is passed through the aortic coarctation and positioned in the ascending aorta. In a tight or tortuous coarctation, it is preferable to cross the lesion with a straight tipped guidewire from below. Adequate angiography and accurate measurements are even more important than for simple balloon dilation. Small errors may lead to selection of a wrong balloon size and stent and therefore increase the complication rate significantly. The length of the stent is based on the length of the hypoplastic segment, typically from the left subclavian artery or the left common carotid artery depending on previous surgical technique and/or site of coarctation to about 15 mm beyond the site of the coarctation.

To reduce the small risk of vascular complications with bare stents, it is reasonable to test the compliance of the coarctation lesion. A balloon of similar size or even bigger than the intended stent is inflated at low pressure. This maneuver is intended as a diagnostic measure, not as an angioplasty prior to stent placement [33]. If there is a significant residual waist on the balloon, a slightly smaller balloon is chosen for stenting

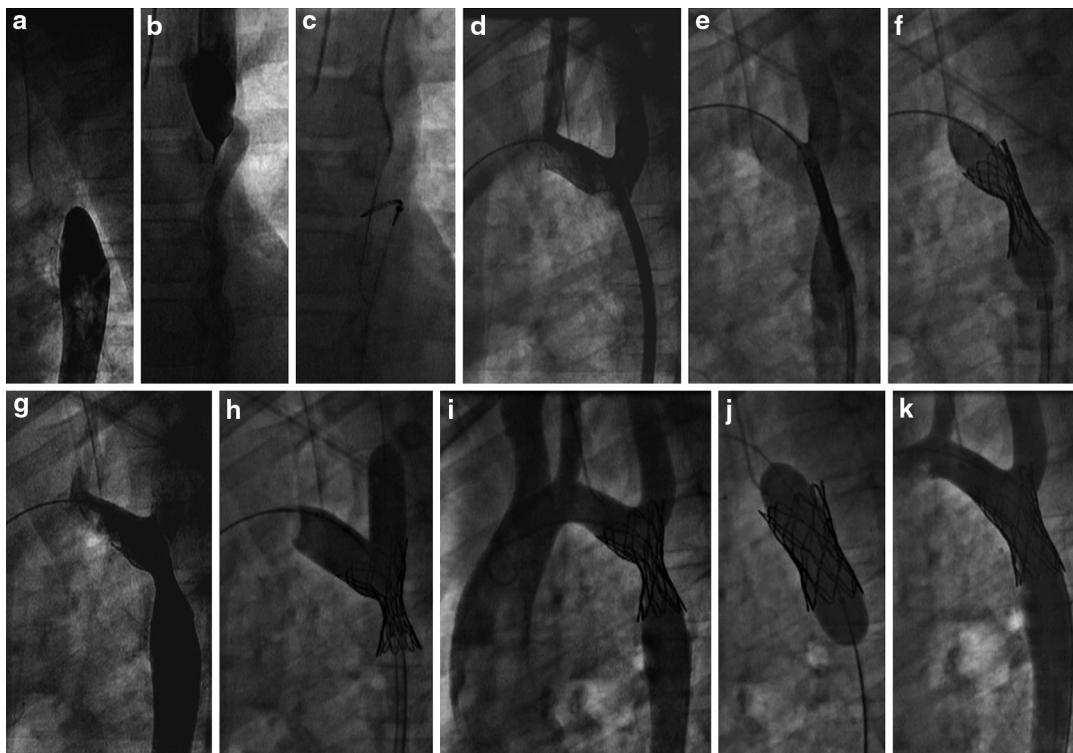


Fig. 64.2 Stenting of near atresia of the aortic arch. Ten-year-old boy with near atresia of the aorta: (a) Aortogram showing no retrograde flow. (b) Aortogram in distal isthmus showing tiny passage of contrast to thoracic aorta. (c) Snaring of 0.014" coronary wire. (d) After arterio-arterial circuit was made, retrograde passage of 11 F sheath and hand injection of contrast through the sheath. (e) 34 mm CCP stent in position on 14 mm BIB balloon. The large origin of left subclavian artery is partly covered but remains accessible after deployment of the stent. (f) Hand

inflation of balloon with subtotal opening of stent sufficient for stent anchoring. (g) Flaring of upper end of stent with 10 and 14 mm balloon to open passage to the left subclavian artery and appose the stent maximally to the wall. (h) Stent nicely apposed to the vessel wall, thereby sealing the zone of expected vessel tear at the end of first procedure. (i) After 2 months, dilation with 12 mm high-pressure balloon at 10 atm. (j) Final result with no residual gradient and excellent patency of stent

and complete expansion of the stent is postponed until a second catheterization 2–6 months later. However, a test interrogation of the coarctation site is usually not indicated if a covered stent is used.

The maximum balloon diameter on which the stent is mounted is based on either the transverse or the distal arch diameter, whichever is the larger, and on occasions 1–2 mm larger. A long Mullins sheath is passed over the 0.035" stiff exchange guidewire. The sheath size ranges between 10 and 14 Fr and is generally 2–3 Fr larger than that required for introduction of the balloon catheter alone (Table 64.1). The stent is manually crimped tight on to the selected

balloon, so as to ensure that it does not slip off the balloon. To facilitate introduction of the stent/balloon assembly through the diaphragm of the sheath, to prevent the stent from slipping off the balloon, and to protect the covering of the stent from being removed inadvertently, a cutoff short sheath tubing of similar size and sufficient length or a dedicated introducer is placed over the stent.

The stent/balloon assembly is advanced through the long sheath and positioned across the site of the coarctation. Optimal positioning is confirmed by small hand injections of contrast through the side arm of the Mullins sheath. Alternatively, injections can be made through a second catheter placed in the transverse aortic arch.

While maintaining the balloon catheter and guidewire position, the Mullins sheath is withdrawn to expose the stent/balloon assembly in position at the site of the coarctation. Keeping the sheath just below the balloon will enhance the stability during inflation. Care must be taken to withdraw the sheath sufficiently below the balloon to allow the balloon inflation in an unrestrained manner. Failure to do so may cause the balloon/stent assembly to move during inflation, or milk the stent off the balloon because of asymmetric inflation. Rapid right ventricular pacing is used if desired. The balloon is initially inflated slowly to allow the shoulders of the balloon to distend and to immobilize the stent on the balloon, and then faster inflation is performed until the stent is anchored at the stenosis site with both ends of the stent widely open and the stenosis sufficiently relieved, or the maximal balloon pressure reached. Once the stent is deployed, the balloon is deflated and pacing is stopped. If a BIB balloon is used, the inner balloon is inflated first, followed by the outer balloon, and for fast deflation, both balloons can be deflated simultaneously. A bare stent can be expanded to the diameter of the normal vessel at either side of the coarctation; however, in case of a tight coarctation, an undersized balloon should be chosen or the balloon not fully expanded in order to reduce the likelihood of aortic wall damage. After deflation, the balloon is withdrawn carefully so as not to dislodge the stent. The gradient across the stent is then measured and an aortogram is repeated to exclude dissection or aneurysm formation.

Further dilation with a larger balloon is performed in some cases until satisfactory relief of the stenotic waist is obtained. Flaring of the ends of a bare stent to achieve contact with the aortic wall at all points is not usually performed. In contrast, flaring of the ends of a covered stent allows adhesion and sealing of the wall, which will prevent a tear from creating an aneurysm or extravasation of contrast. In complex lesions in small patients, it is not uncommon to use one low-profile balloon to deliver a stent across a stenosis through a small sheath, another larger balloon to flare and appose the ends of the covered stent to the wall to obtain maximal sealing, and one or

more high-pressure noncompliant balloons to progressively dilate the stenotic region.

Patients are usually discharged the day after the procedure and reevaluated clinically and echocardiographically 4 weeks, 6 months, and 1 year after the procedure. Spiral CT scanning is performed 4–6 weeks after the intervention to exclude aneurysm formation, dissection, and stent thrombosis.

Anticoagulation and Antiplatelet: Follow-Up

A large diameter stent with high flow is unlikely to thrombose. Many interventionalists neutralize the heparin with protamine at the end of the procedure when removing the sheath and give no further antithrombotic treatment. Several other regimes have been advocated, but no trial has been reported to prove the need or superiority of any anticoagulation or antiplatelet protocol.

Re-catheterization is performed only if there is clinical evidence of recoarctation, residual arterial hypertension, or CT evidence of aneurysm formation. In patients in whom the stent was initially intentionally underinflated, elective re-catheterization is performed after 2–6 months to relieve residual stenosis.

Bare or Covered Stents

An aneurysm or aortic wall rupture can occur unexpectedly; therefore, the procedure with a covered stent is safer than with a bare stent, as the lesion is automatically contained. A covered stent offers a bigger margin of safety to expand the stent more at the initial procedure and much more at subsequent interventions, if indicated. The use of covered stents is necessary to safely treat patients with atretic or severely hypoplastic segments, as a transmural tear may occur. Not all problems are abolished as tears at the edges of the covered stent [34] or at a distance, bleeding from the vessel wall or the side vessels such as intercostal arteries may still occur, but these complications are usually less dramatic. Stenting techniques are slightly different

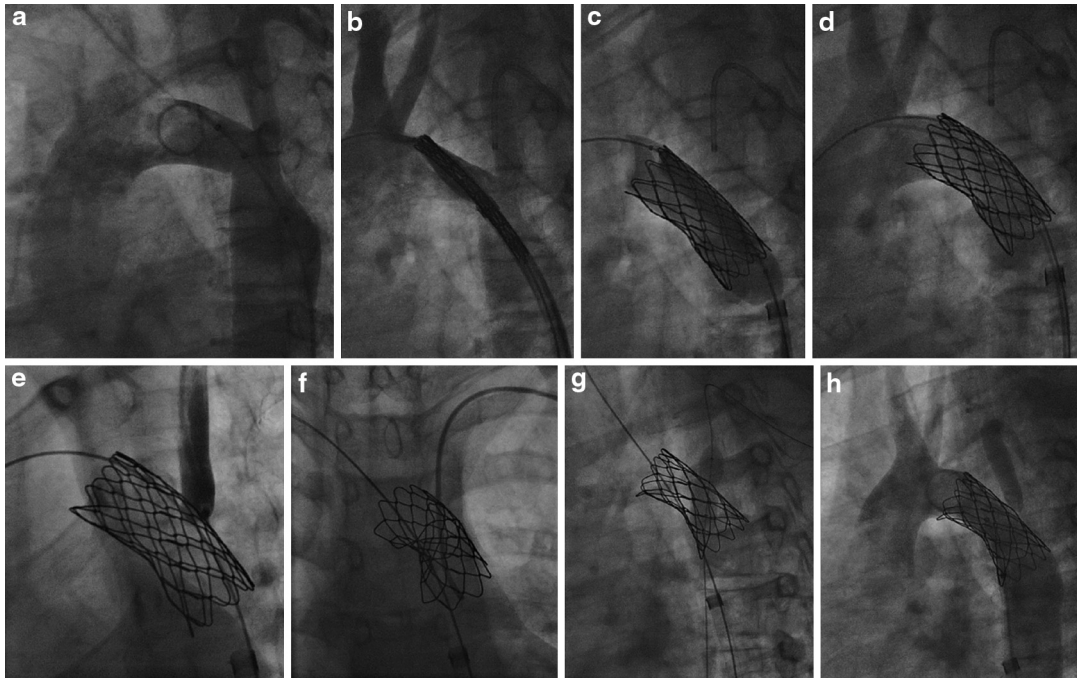


Fig. 64.3 Retrograde puncture of covered stent to establish antegrade flow in the neck vessel. A 14-year-old girl with postoperative residual coarctation at the distal aortic arch: (a) Aortogram showing complex recoarctation; residual hypoplasia of distal arch at the origin of the left subclavian artery. (b) 45 mm CCP[®] stent mounted on a 16 mm BIB[®] balloon through a 13 Fr Mullins sheath. Hand injection through side arm of the sheath demonstrates correct position of the stent. A 4 F end-hole catheter is positioned in the left subclavian artery to perform

the retrograde puncture. (c) Stent is deployed with the 16 mm BIB[®] balloon. (d) Aortogram through sheath demonstrates correct deployment of the stent. (e) Injection in the left subclavian artery through the 4 F catheter. (f) After retrograde perforation with stiff end of 0.035" guidewire, the 4 Fr catheter is advanced into the thoracic aorta. (g) Balloon dilation of stent covering at the origin of the left subclavian artery with a 10 mm balloon. (h) Final result with no gradient across the aortic arch and antegrade flow into the left subclavian artery

when using covered stents compared with bare stents. Balloon interrogation prior to stenting is rarely performed and the stent is fully approximated against the wall at deployment in order to obtain maximal sealing around the hypoplastic segment. The combination of a CCP stent with a slightly oversized BIB balloon allows the stent to be apposed fully to the aortic wall at the site of the coarctation. Progressive dilation of the coarctation site is performed either immediately or at a later procedure, depending on the operator's estimation on how well the expected tear will be sealed off. The ends of the stent can be flared into the side vessels such as the subclavian or left carotid artery, if required.

Experience with stent grafts for thoracic aortic aneurysms suggests that the origin of the left

subclavian artery can be covered with a covered stent without any acute effects although it is preferable to avoid this. However, arm claudication can rarely occur. Additional techniques such as retrograde perforation [35] and double wire technique [36] may therefore be required to avoid excluding side vessels. *Retrograde perforation* is an elegant technique which allows stenting of a narrowing at or near the origin of a side vessel such as the subclavian artery and the carotid artery. The stent is deployed in the arch and the covering is perforated retrogradely from the excluded vessel that has been covered, thereby restoring antegrade flow (Fig. 64.3).

The double wire technique allows a stent to be delivered across a stenosis or aneurysm just distal to a side branch (Figs. 64.4, 64.5 and 64.6).

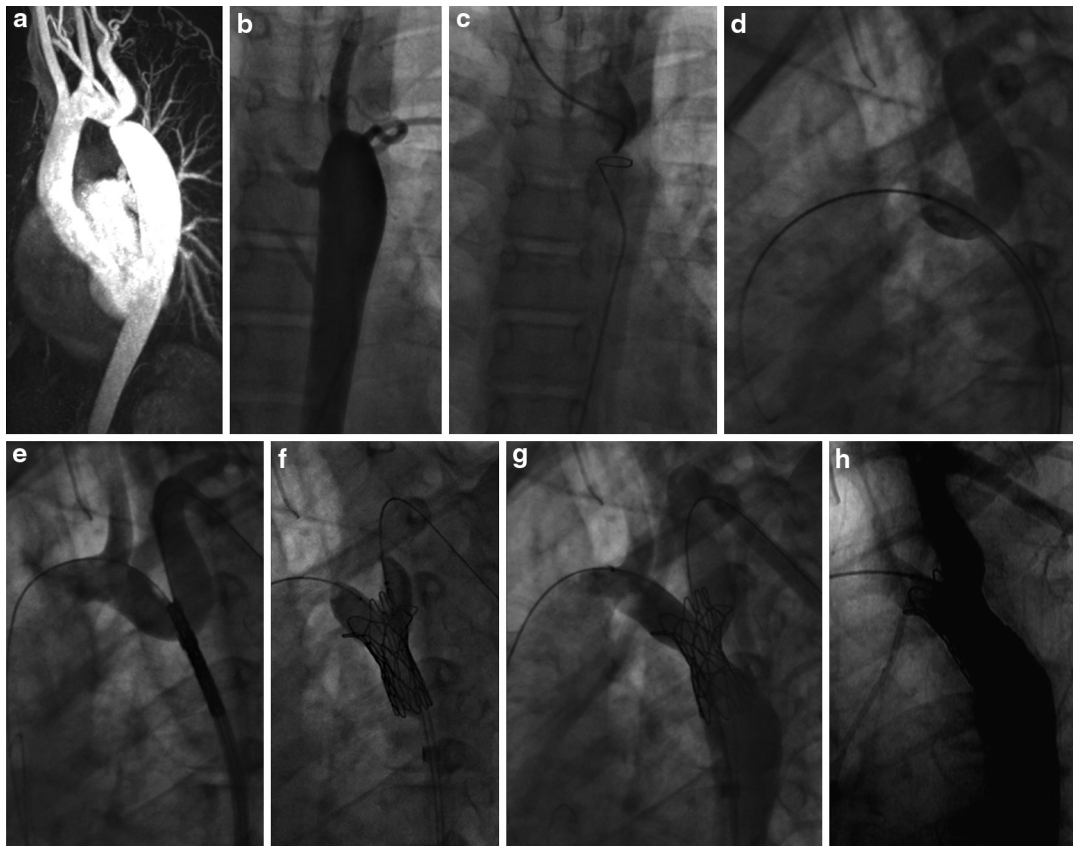


Fig. 64.4 Double balloon technique. A 13-year-old girl presented with a near aortic arch atresia. (a) MR demonstrates the near atresia. The left subclavian artery originates just proximal to the coarcted segment. (b) Aortogram in the descending aorta demonstrates large collateral vessels, but no connection with the arch. (c) Antegrade passage from the right brachial artery as the angle from the left subclavian artery was assessed to be too acute. A coronary guidewire is captured with a 10 mm snare in the descending aorta to form an arterio-arterial circuit. (d) Aortogram in the distal arch. Note a 0.035" guidewire placed in the ascending aorta. (e) After

predilation with a 3 mm balloon, a 11 Fr long Mullins sheath is positioned in the distal arch. Two 0.025" guidewires are positioned in the ascending aorta and left subclavian artery. A 34 mm CCP® stent mounted on a 12 and 10 mm Tyshak® balloon is positioned as cranial as possible. (f) The stent is opened and delivered by gentle inflation of both balloons. Dilation of the coarctation site at this point is no issue. (g) The stent is apposed to the wall thereby sealing the expected zone of vessel tear at the end of first procedure (h) 2 months later, the stent was expanded with a 14 mm high-pressure balloon and there was no residual gradient

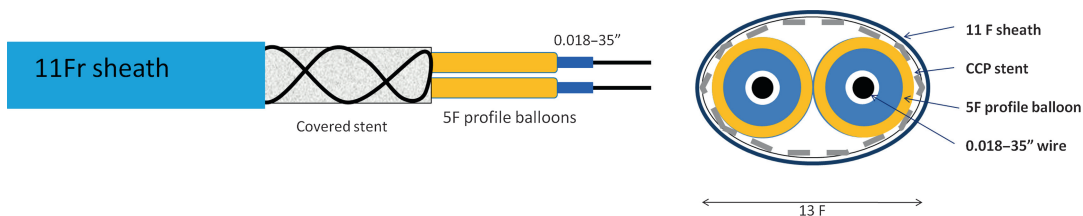


Fig. 64.5 Double balloon technique: sheath size. The smallest sheath must be determined for any selection of balloons and stent. A CCP stent mounted on two balloons

each with a 5 Fr profile has an oval shape and can be delivered through an 11 Fr Mullins sheath

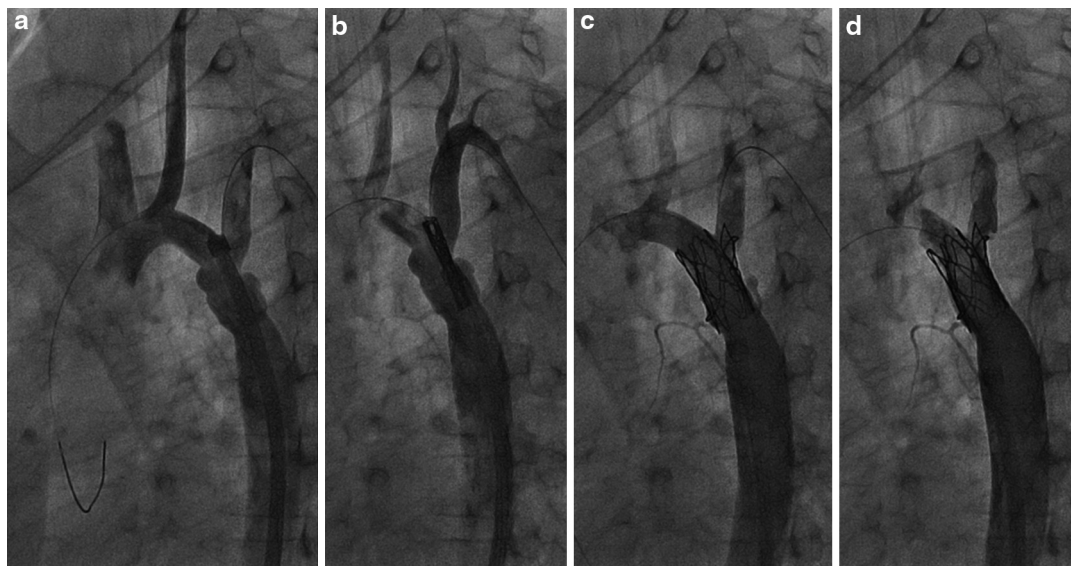


Fig. 64.6 Double balloon technique to exclude post-dilation aneurysm. A 10-year-old boy weighing 28 kg presented 5 years after balloon dilation of coarctation with a complex aneurysm at the dilation site, involving the origin of the left subclavian artery. (a) Aortogram through a 11 Fr sheath after two 0.018" guidewires have

been positioned (b) 22 mm CCP stent, mounted on two 10 mm Tyshak[®] balloons, is positioned as cranially as possible to cover the aneurysm. (c) The stent is deployed but there is still a small opening into the aneurysm distally. (d) After further expansion with a 14 mm balloon, the stent seals off the aneurysm completely

The second wire maintains access to the side vessel after deployment of the stent and allows flaring of the stent into the side branch. In order to reduce the required sheath size, the lowest profile balloons to get the stent anchored are selected, without the aim of dilating the lesion at delivery. After anchoring the stent, it is subsequently expanded as needed. Once balloons are selected, sheath size is determined by bench testing. Theoretically, one would add the profile of the two balloons to the necessary upsizing for a bare or covered stent. However, the whole unit becomes oval in shape instead of the normal round configuration, allowing a decrease in the final sheath size from the mathematical prediction (Fig. 64.5).

If sheath size is critical, both CCP and V12 covered stents can be delivered through a 10 Fr sheath, followed by additional dilation with a larger balloon as indicated.

In many centers, a gradual shift from using bare stents to using covered stents has occurred due to the additional therapeutic and safety margins. Disadvantages of covered stents are the

slightly bigger profile and potential hemodynamic problems if the stent migrates inadvertently. This complication can be avoided by correct technique and attention to detail.

Measures of Success

The main measures of success of treating coarctation should be abolition of the gradient, control of blood pressure, and absence of complications from the treatment. While treatment with balloon-expandable endovascular stents is a technically challenging procedure, it is an extremely successful one [17, 22, 37–39]. A multicenter retrospective series of 588 procedures performed between 1989 and 2005 was conducted by the Congenital Cardiovascular Interventional Study Consortium (CCISC) [40]. Of the 588 procedures, 580 (98.6 %) were successful in reducing the gradient to less than 20 mmHg or increasing the coarctation to descending aortic diameter ratio to at least 0.8. Two patients developed an aortic dissection with

rupture and the procedures had to be terminated and emergency surgery undertaken with a bad outcome. In retrospect, such complications were probably avoidable and treatable with the use of covered stents.

In the adult population, the initial gradient across the coarctation may not reflect its severity as there may be extensive collateral vessels decompressing the aorta proximal to the stenosis. Resolution of hypertension cannot necessarily be used as a measure of efficacy because the incidence of hypertension may be masked by antihypertensive treatment. Some adult patients without residual stenosis at the coarctation site will continue to be hypertensive with persistent abnormal endothelium [41, 42]. However, control of their blood pressure usually becomes easier after stenting. Stenting appears to be effective in reducing resting blood pressure to normal levels in the majority of children and adults.

Little information is available on how stenting affects exercise tolerance or how well the stented coarctation segment responds to the increased cardiac output in pregnancy.

Complications

Some complications of coarctation stenting have already been discussed. In general, they can be classified into technical, aortic wall or peripheral vascular complications, or post-procedural hypertension and pain.

Technical complications include stent migration on the balloon in the sheath, during deployment on the balloon, migration after deployment, stent fracture, balloon rupture, and covering of the brachiocephalic vessels [43]. While passing the stent/balloon assembly through the valve or the sheath, the stent may migrate off the balloon; radiopaque markers on the balloon allow confirmation of correct position of the stent on the balloon before withdrawal of the sheath to uncover the stent in the aorta. If the stent has moved, the stent-balloon sheath can be removed leaving the guidewire in place, remove the stent from the front of the sheath and start the procedure again.

Stent migration off the balloon during inflation can occur if the balloon is inflated asymmetrically. This can be avoided by minimal inflation of the balloon before introduction through the sheath, creating small shoulders on both sides of the stent. This is relatively easily done with an indeflator. During stent deployment, balloon inflation should be started slowly, allowing both shoulders of the balloon to expand, thereby immobilizing the stent on the balloon. Stent migration can be avoided further by using a BIB balloon, especially when using the bigger sized balloons of >15 mm, the inner balloon is typically within the stent and cannot milk the stent off the balloon and the outer balloon will inflate symmetrically after the inner balloon has been inflated. During or after deployment, the stent may migrate more proximally or distally. Often the stent can be recaptured with a balloon and repositioned. If it cannot be repositioned safely within the coarctation, it should be expanded in the safest location available, away from side branches if possible. *Balloon rupture* may be avoided by using an appropriate balloon for a given stent. Stents with sharp edges require thicker, puncture-resistant balloons. Balloon rupture occurred in 13/588 (2.2 %) of cases in the CCISC cohort predominantly when using older stents such as the now-abandoned Palmaz 8-series stents. Balloon rupture may result in other complications involving the aortic wall, or embolization of balloon fragments, and if the balloon ruptures prior to full expansion, it will carry a high risk of stent migration.

Whether stent placement over the *origin of the brachiocephalic vessels* constitutes a complication is debatable. There have been no demonstrated harmful sequelae from doing so, except at redilation (see below [44]). A late *stent fracture* may occur at the transition of the mobile segment of the aortic arch to the fixed retropleural thoracic aorta. Currently, stent fractures may occur in stents with thinner metal, e.g., Genesis and Valeo stents, when expanded to larger diameters.

Aortic wall complications at or around the site of the coarctation include *intimal tears, dissection, aneurysm formation, and rupture* either

within the stent or at the edges or at a distance [45–47]. Vascular complications are more prone to develop in patients with connective tissue disease such as Turner syndrome [48]. Most of these complications can be treated, or are better avoided, by using covered stents [49, 50]. The general rule of “it is easier to stay out of trouble than get out of trouble” certainly applies to these situations.

It is important to have large diameter covered stents available for use in emergency situations as the covered CP stent can be dilated up to 24 mm and Atrium stent up to 20–22 mm, but for some emergencies, larger self-expanding excluder stent grafts (from Boston Scientific, Gore, Medtronic) should be available.

Aortic aneurysm is infrequently encountered, but it may be a harbinger of aortic rupture and is therefore a potentially dangerous complication. It may be seen at the time of the procedure or on follow-up. If a large or growing aneurysm occurs at the time of the stent placement, it must be excluded with a covered stent to prevent progression and possible rupture [51–53].

Peripheral vascular complications include cerebrovascular accidents, peripheral emboli, and injury to access vessels. Neurologic events including cerebrovascular accidents occurred in the CCISC group in 6/588 procedures. Adequate anticoagulation during the procedure is essential as the head and neck vessels are crossed with wires for a prolonged time and long sheaths are used where clots may form. Horner syndrome was reported due to a carotid artery dissection by the guidewire [30].

Significant femoral vessel injury was reported in the CCISC study in 15/588 (2.6 %) procedures. One patient had placement of the arterial sheath above the inguinal ligament and developed a retroperitoneal hematoma. Vessel thrombosis is more frequent in small children. It is current practice to institute heparin therapy for 24 h when there is loss of pulse after catheterization. If the pulse has not returned after 24 h, or if the viability of the leg is a concern at any point, thrombolytic therapy or surgery may be indicated.

Post-procedural rebound hypertension is sometimes observed in adult patients

immediately after the procedure. Patients with systolic blood pressures greater than 99th centile for age should be monitored carefully, and infusions of nitroprusside or esmolol or both should be used, if there is severe rebound hypertension. These patients can generally be switched to oral antihypertensive medications within 24 h after the procedure.

Thoracic pain and abdominal discomfort can occur early after the procedure. This pain may only become evident when the analgesics from the anesthesia fade away. Thoracic pain remains an alarming symptom, so dissection, aneurysm formation, bleeding from the aorta, or torn intercostal arteries must be excluded by observing peripheral pulses and assessing by echocardiography and CT scan. Such pain is most likely due to stretching of the aorta and requires adequate analgesia in the form of opiates and is usually relieved after some hours. Some adult patients may complain of abdominal discomfort early after the procedure because better pulsatile flow may cause bowel irritability within the first few hours after the intervention.

Special Situations: Bail-Out Stenting in Premature Babies

A coarctation in newborns is typically treated by surgery. However, there may be occasions when a clinician may prefer to defer the surgery. These include extreme prematurity, a critically ill neonate with multi-organ failure shock, or complex syndromic patients. Stenting a coarctation may be performed as an emergency in such patients and may acutely improve the newborn, deferring surgery to a later period after adequate weight gain or hemodynamic stability. This strategy compares favorably to a surgical treatment strategy, when applied in critically ill or vulnerable newborns [54].

The technique in these premature infants is slightly different from that previously described. Puncture of the femoral artery is performed with a 21 gauge needle allowing a 0.014" wire to be introduced into the artery. A 4 Fr smooth tapered introducer sheath is placed in the artery. A 4 Fr

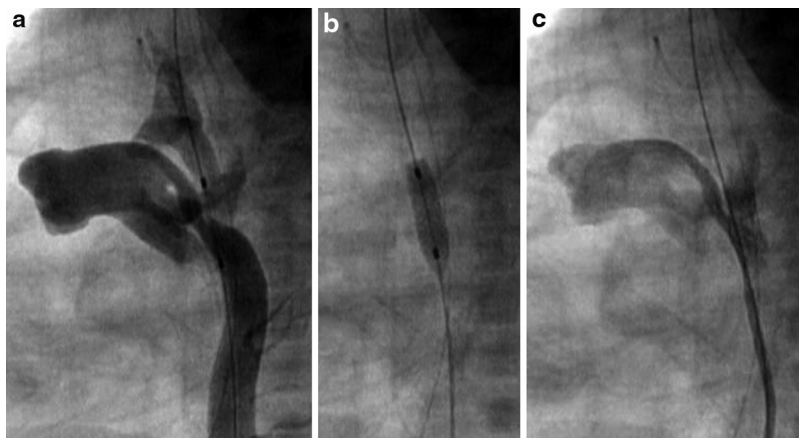


Fig. 64.7 Bail-out stenting in a premature infant. A 1,500 g infant presented with critical aortic coarctation. (a) Retrograde aortogram by hand injection of layered contrast-saline through a short 4 F sheath. A 4/8 mm coronary stent is positioned for deployment. Points of

reference are the cranial end just beyond take-off of left subclavian artery and caudal end beyond coarctation site within the thoracic aorta. (b) Inflated balloon has expanded the stent. (c) Hand injection of contrast through 4 F catheter to confirm adequate position of stent

end-hole vertebral catheter is advanced up to the coarctation site where a small volume of 1 cc of contrast is injected by hand. The coarctation is crossed using an atraumatic 0.014" coronary wire. If the isthmus cannot be entered retrogradely, a transvenous antegrade approach may be used. Another hand injection is performed in the aortic arch to delineate the anatomy and the origin of the left subclavian artery. A low-profile, premounted coronary stent is chosen on the basis of the pre-catheter echocardiographic measurements as well as angiography. The stent should ideally cover the arch from just distal to the origin of the left subclavian artery up to and beyond the coarctation site, the typical length being 8–12 mm. The stent diameter should equal the aortic arch diameter, which is usually about 3–5 mm. Such diameter allows a significant increase in size at the coarctation site, without risk of vessel tear because “fetal tissue” may allow significant stretch.

The stent is passed “unprotected” through the valve of the introducer sheath. Stent position is checked with retrograde aortogram hand injections through the short femoral sheath. A useful method is using sequential gentle aspiration of 5 cc of saline into a 10 cc syringe held vertically, followed by aspiration of 1 cc of contrast,

keeping contrast and saline layered (Fig. 64.7). The stent is deployed using an inflator device at a pressure recommended by the manufacturer. In premature babies and neonates with primary coarctation, the prostaglandin E-1 infusion is stopped immediately after deployment of the stent. Stent position is assessed with an aortogram through the 4 F catheter placed just below the stent. Such stents usually produce a satisfactory result for several weeks to months. In very small premature babies below 1,000 g weight, vascular complications may be avoided by accessing the aorta directly during a hybrid procedure. After sternotomy, a 4 F sheath is inserted in the ascending aorta, allowing the stenting procedure to be performed. Such an approach also allows for closure of the patent arterial duct [55] (Fig. 64.8).

The timing of surgical removal of the stent may vary. After some days when the neonate has recovered sufficiently from the initial cardiogenic shock, or after some weeks or months when adequate body weight has been reached to perform surgery safely, or when additional surgery is planned. If during follow-up more time is required, any coronary stent can be further dilated up to 5 mm as this also reduces the stent length if this were an issue for safe resection.

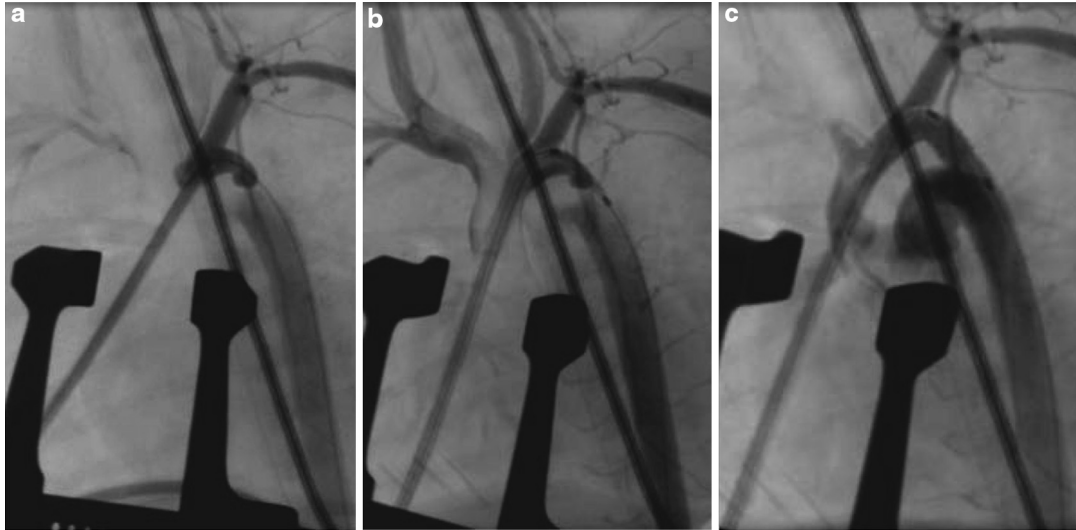


Fig. 64.8 Antegrade bail-out stenting in 850 g premature infant through sternotomy. An 850 g infant presented with critical aortic coarctation. (a) After sternotomy, a 4 Fr sheath was inserted in the ascending aorta through a purse string. Aortogram is performed by hand injection. (b) A 3/8 mm coronary stent is positioned for deployment. Points of reference are cranial end just beyond take-off of

the left subclavian artery and caudal end beyond coarctation site within the descending aorta. (c) Hand injection of small volume of contrast through 4 F catheter to confirm correct position of stent. The duct was then clipped surgically and the sternum closed. The stent was surgically removed 4 months later.

Dilation: Redilation

Staged dilation, in which stents are expanded to a diameter less than the adjacent aorta and redilated a few months later, may overcome the possible risks of excessive wall damage. A controlled injury is allowed time for healing of the arterial wall before full expansion is attempted. However, even with this approach, aneurysm formation may occur.

If stents are implanted in smaller patients, somatic growth of the patient will require further redilation. In this age group, stenting should be reserved for exceptional clinical indications rather than routine use because excellent surgical results can be obtained with extended arch repair.

Redilation of a stent is generally a straightforward procedure. However, such procedure may still be associated with complications. Within a bare stent, an aneurysm may develop and embolization of intimal peel may occur; dissection and aneurysms may develop at the stent ends, but also at a distance. Most stents shorten when dilated to larger diameter, causing

longitudinal stress on the vessel wall. Theoretically, it is safer to dilate gradually in small steps and with short balloons. An excessive shoulder of a big long balloon may tear the vessel wall from the stent before effective dilation. Such complication has been reported with late dissection of the thoracic aorta and fatal outcome [48]. An aortogram should therefore be performed at least at the end of any procedure, and if such complication is observed or suspected, implantation of an additional covered stent may be lifesaving.

When using covered stents, the operator should aim for adequate apposition of the stent to the wall, thereby allowing maximal adherence of the stent around the narrowed segment. Such safety zone will seal the expected tear at further dilation.

Arch Atresia

A long standing critical coarctation may occlude completely, creating an atretic segment by the

time treatment is initiated. The pre-catheterization evaluation should attempt to determine whether there is critical stenosis or atresia. Careful Doppler examination may show a tiny connection on color flow or a typical sawtooth pattern. If atresia or critical stenosis is expected, access through both the brachial/radial and femoral arteries must be obtained. Angiography is performed proximal and distal to the interrupted segment. In some patients, a minute connection may be found when actively looked for. This can be crossed with the use of a 0.014" coronary guidewire or a 0.018" guidewire in a 2 Fr tracking microcatheter system [56]. When no connection is found, recanalization may be performed by puncturing with a stiff end of a guidewire of 0.035" caliber or a 0.014" guidewire or a Brockenbrough needle or by radiofrequency perforation using a Nykanen 0.024" wire through a coaxial system (Baylis Medcomp, Montreal, Canada) or a PT2 coronary wire through a 2 Fr Progreat catheter passed through a 4 Fr catheter [57]. The anatomy of each patient will determine how the atretic portion is best crossed. Usually, this is from the arch to the thoracic aorta, with an opened snare in the descending aorta as target. Once the guidewire is snared, an arterio-arterial circuit is established. Predilation may be required to allow crossing the defect with the delivery sheath. A covered stent is implanted with sufficient overlap proximal and distal to the atretic segment. Full stent expansion may be performed a few weeks after the initial stenting procedure.

Stenting of Transverse Aortic Arch

Many patients with a coarctation may have associated hypoplasia of the transverse arch, which can leave a residual gradient. The surgical treatment option for transverse arch hypoplasia is to perform an extended arch repair, which frequently requires cardiopulmonary bypass and is not risk free, or insertion of a bypass graft.

Percutaneous treatment of arch hypoplasia is slightly different from coarctation treatment [58, 59]. Balloon dilation is likely to be

unsuccessful, and so stenting may be necessary. The margin between a therapeutic tear and a catastrophic rupture is very small. The adventitia is adherent to the media in the transverse arch, which is not the case in a typical coarctation where the adventitia "jumps" from the isthmus over the coarctation to the thoracic aorta. Therefore, when stenting the transverse arch, the aim is to stretch the wall without creating a tear. In the proximal transverse arch, bare metal stents are almost exclusively used, while in the distal arch, a covered stent may be used. Such stents may be positioned across the origin of the left subclavian artery and exclude it. This can then be reopened easily by retrograde perforation of the covering and balloon dilation of the orifice (Fig. 64.3).

An ascending angiogram is performed in two planes perpendicular to each other with the aim of defining the arch anatomy as well as the origins of the innominate, the left common carotid, and the left subclavian arteries. Balloon interrogation of the hypoplastic segments of the arch with a low pressure, mildly oversized balloon such as Tyshak balloon (NuMed) is essential. The stretchability of the segments can be assessed, which determines the balloon size and the desired stent conformation during deployment (Fig. 64.9). This technique allows the operator to avoid deployment of an undersized stent which will anchor itself insufficiently in the wall and possibly migrate, or to tear a narrow, unstretchable segment. The balloon diameter is chosen based on the largest stretched diameter of the aortic segment to be stented. The stent needs to be about 3–5 mm longer than the distance between the origins of the vessels where the aorta is to be stented. Stents partially protruding over the origin of the left common carotid artery can be flared into the vessel, whereas struts covering the origin of the left subclavian artery are of less concern. During inflation, the stent will shorten asymmetrically, typically around the point where the stent touches and anchors itself in the aortic wall.

Angiograms are performed through the side arm of the Mullins sheath to check for accurate positioning of the stent. The Mullins sheath is withdrawn while keeping the stent/balloon

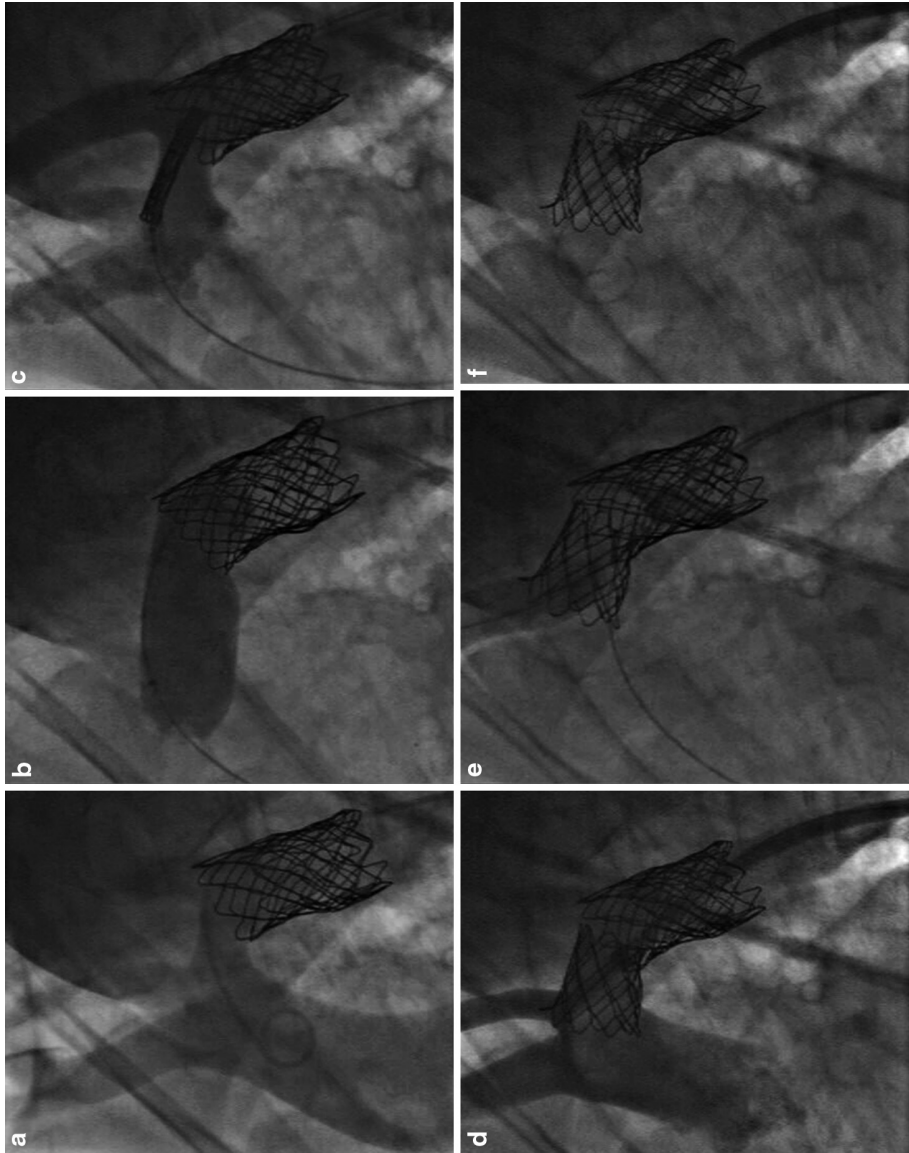


Fig. 64.9 Stenting of the hypoplastic transverse arch. A 19-year-old boy with previous stenting of coarctation and residual hypertension: (a) Aortogram shows 12 mm diameter hypoplasia of distal arch between the left common carotid and the left subclavian arteries. There was a gradient of 15 mmHg. Coarctation was previously stented with 20 mm diameter stent. (b) Low-pressure balloon interrogation with 20 mm Tyshak[®] balloon to determine compliance of hypoplastic segment. (c) Deployment of 28 mm bare CP stent on 20 mm BIB[®] balloon at low pressure through 13 F sheath. (d) Stent in transverse arch. (e) Flaring of stent into left carotid artery with 16 mm balloon. (f) Final result with no residual gradient

assembly in position so as to expose the stent. The balloon is inflated manually at low pressure. A BIB balloon allows deploying the stent as accurately as possible. The expansion of the stent is usually symmetrical during inflation of the inner balloon, and asymmetric shortening only occurs when the stent anchors itself in the aortic wall. The stent is apposed to the wall under low pressure, aiming to stretch and not tear the wall. On balloon inflation, the stent shortens and so may clear off the origin of the carotid artery, or the end of the stent can be flared into it.

Middle Aortic Syndrome

Middle aortic syndrome is an uncommon lesion presenting with physical signs of coarctation of the aorta, hypertension, renal insufficiency, and/or mesenteric ischemia. The etiologies are multiple, but Takayasu's arteritis is a leading cause. Variable involvement of diverse systemic arterial systems requires individualized management strategies. The aortic wall becomes very thick and pressure resistant, and balloon dilation alone yields poor results. Stenting using high-pressure balloons is required, with a significant risk of vessel tear and stent thrombosis [60]!

What if a Residual Gradient Persists After Percutaneous Stenting

Despite percutaneous stenting and an optimum result, a significant residual gradient may persist due to residual hypoplasia of the aorta, which cannot be dilated safely, or due to angulation in a high cervical Gothic arch, restrictive stents, or conduits. Surgery in an arch with gradients at different levels can be difficult, even more so when some segments are stented. Adequate gradient relief can then be obtained with an extra-anatomic bypass [61]. Two types of approaches can be applied. A repair through a median sternotomy consisting of creating a "right arch," with the insertion of a bypass from the right lateral wall of the ascending aorta, routed around the right margin of the heart, to the supra-celiac

abdominal aorta. A second approach is through a left thoracotomy with interposition of a graft between the ascending and descending aorta or interposition of a graft between the left subclavian artery and the descending thoracic aorta.

True Aneurysms

A true aneurysm consists of dilation of all layers of the vessel wall. Small aneurysms can be treated very effectively with a covered stent as described above. However, several treatment strategies, both surgical and interventional, can be complicated later with large and long thoracic aneurysm formation. Such aneurysms are associated with a high rate of rupture within 15 years after detection [62]. Traditionally, these aneurysms are treated by repeat surgery including interposition graft placement under cardiopulmonary bypass, hypothermic circulatory arrest, or other methods of distal circulatory support. Post-operative mortality may be as high as 13 % and morbidity may include paresis of the left recurrent laryngeal nerve and bleeding requiring repeat thoracotomy. Thoracic endovascular aneurysm repair (TEVAR) has emerged as a minimally invasive alternative for repeat surgery after coarctation repair [63–65]. These techniques however involve self-expanding systems requiring large sheaths of 22–24 Fr, which necessitate a surgical shutdown.

Pre-interventional imaging is usually performed with magnetic resonance and/or computed tomography imaging, including 3-D reconstructions. The diameter of a thoracic aortic stent graft should be oversized by 10–15 % compared with the nominal diameter of the proximal and distal landing zones. The length of the stent graft should include the length of the aneurysm and the length of the proximal and distal landing zones, which are at least 2 cm each. Typical contraindications for conventional TEVAR are too small proximal luminal diameter, too short length of the landing zone, and an acute angle of the thoracic arch such as Gothic arch. These cases should be considered for open surgical or hybrid repair.

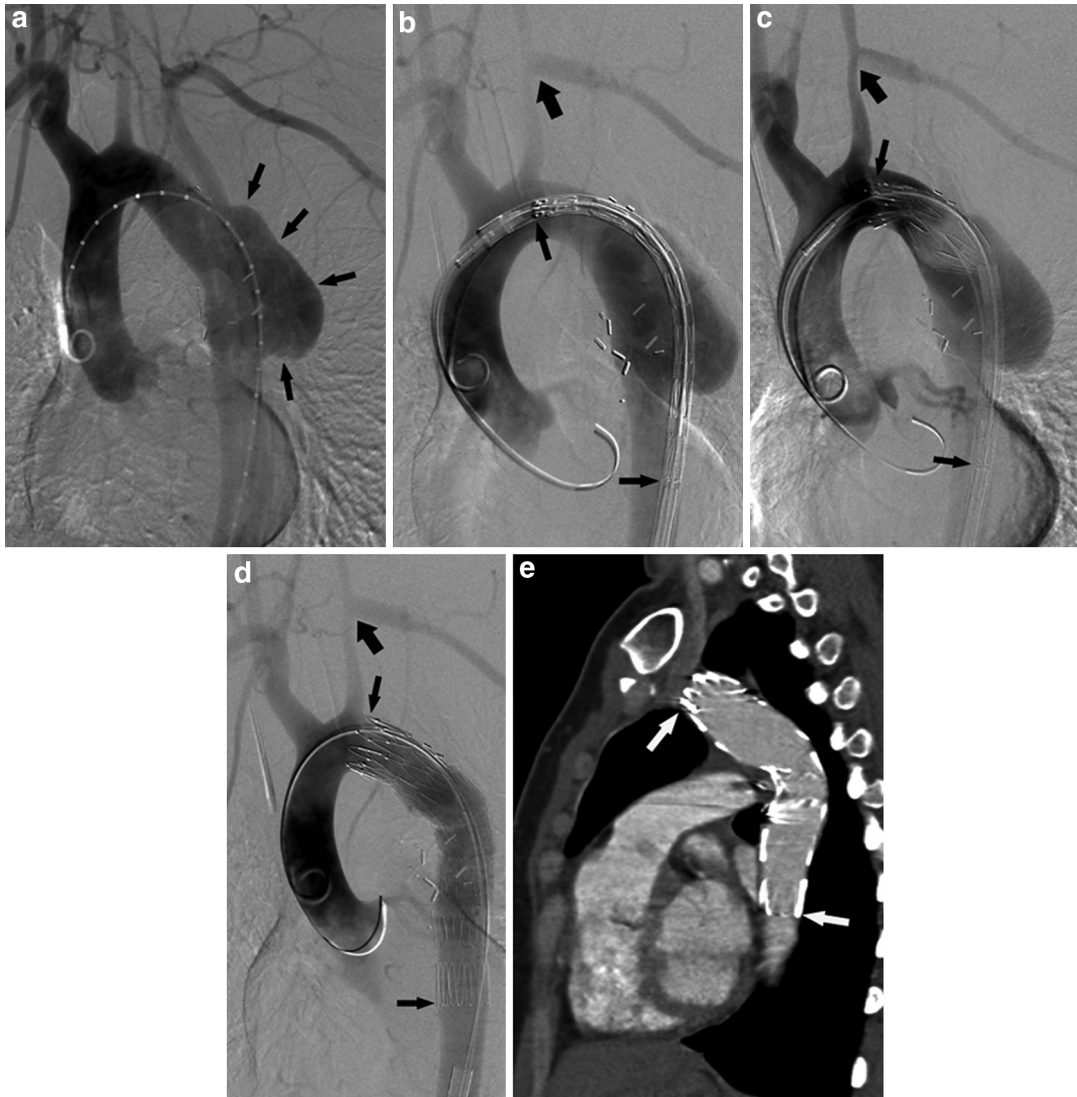


Fig. 64.10 Exclusion of aneurysm by stent graft. A 32-year-old woman presented with an asymptomatic aortic aneurysm, 28 years after Dacron patch aortic arch repair. (a) Calibrated catheter flush aortography revealed an aneurysm of 50 mm (arrows) distal to the origin of the left subclavian artery. (b–d) Transfemoral insertion of

a Zenith TX2[®] (Cook) stent graft (small arrows) loaded in a 20 F sheath after carotid to subclavian artery transposition (large arrow). (e) Repeat CT scan 6 months after the stent-graft procedure shows the stent graft (white arrows) completely excluding the aneurysmal sac

The stent graft is positioned over the aneurysm and carefully deployed under hypotension induced by rapid right ventricular pacing to reduce the systolic blood pressure to less than 80 mmHg and angiographic and transesophageal echocardiographic imaging control (Fig. 64.10). In case of a small diameter arch and a normal diameter descending thoracic aorta, a reversed

and tapered custom-made device is preferred [66]. Following deployment, the device can be further conformed and approximated using a large, compliant balloon, therefore obtaining optimal proximal and distal sealing.

During the last decade, several small case series, mostly consisting of less than ten patients, have been published [67–69], all demonstrating

that TEVAR is relatively safe and effective for endovascular repair of aneurysms associated with coarctation surgery. Major complications seem to be rare, although perioperative and postoperative mortality may occur [70, 71]. Procedure- and device-related complications encountered after TEVAR for coarctation aneurysms include left-sided arm claudication due to intentional covering of the left subclavian artery, endoleaks, infolding, and collapse of the stent graft, graft infection, stent migration.

Conclusions and Recommendations

Although treatment of coarctation of the aorta with balloon-expandable endovascular stents is technically challenging, it is a relatively safe and extremely effective treatment modality when used carefully in selected patients. The shift from simple balloon angioplasty to implantation of bare stents and eventually covered stents has significantly improved results while decreasing complications. Further research is necessary to determine the incidence of various complications and identify risk factors, allowing refinement of guidelines for even safer and more successful procedures.

With current knowledge and experience, the following recommendations can be made:

- (a) In infants and children less than a year of age, surgery is the treatment of choice for all native coarctation, and balloon angioplasty is the treatment of choice for most recurrent coarctations. In very premature and critically ill neonates, bail-out stenting may be considered, and later surgical stent removal may avoid many complications typically observed in this difficult age group.
- (b) Between the ages of 1 year and the time when the child reaches a weight of 30–35 kg, usually 9–11 years, there is insufficient data to determine whether surgical intervention or balloon angioplasty is preferable for native lesions. Percutaneous treatment usually involves several interventions during growth, while surgical results in a single procedure are very good. Balloon angioplasty is the

treatment of choice for recurrent coarctation in this age group.

- (c) In children weighing more than 35 kg who have not reached adult size yet, it is likely that the treatment of choice for native and recurrent lesions could be endovascular stent placement, as it has been demonstrated that stents can be enlarged safely at a later time to accommodate somatic growth.
- (d) In adolescent and adult patients, stent placement is the treatment of choice for all lesions, whether native or recurrent.
- (e) In many situations, the use of covered stents is emerging as the safer option, especially in adults of advanced age and in patients with known vasculitis or other conditions associated with vasculopathy.
- (f) TEVAR is a good option to treat large thoracic aneurysms.
- (g) All patients require long-term follow-up for timely detection of aortic aneurysms or dilation, as well as arterial hypertension.

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