

Aortic Coarctation: Current Solutions for a Complex Condition

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Aortic coarctation is a frequent and serious clinical entity that decreases life expectancy; without treatment it is associated with high morbidity and mortality.¹ It has an important impact on the clinical, social and working life of the treated or untreated patient, and requires ongoing follow-up, due the threat of immediate or future complications, such as: aortic rupture, acute stroke, arterial hypertension (AHT), and premature coronary heart disease.

The lesion is characterized by thickening of the aortic medial layer with intimal hyperplasia in the posterior wall. Differentiated smooth muscle cells are also found in small numbers and cause a variable blockage to the passage of blood. A large number of pseudocysts are also found on the wall with a high mucin content and without an inflammatory component. It is accompanied by dilatation and poststenotic parietal thickening. The aortic wall presents a variable degree of cystic necrosis of the medial layer that increases with age, until it becomes clearly pathological.² In the newborn, the region affected also presents intimal thickening and abundant smooth muscle cells, possibly related to its genesis.

The anatomy of the lesions is very varied (from just one acute localized coarctation, to more complex lesions with variable hypoplasia of the aortic isthmus and even of the transverse arch) which considerably influences the type of treatment. This should be personalized following a very careful anatomical study.

Surgery has changed the natural history of the disease, from the first intervention in 1945 up to extended end-to-end anastomosis representing one of the best current alternatives. Non-surgical treatment began in

1982, via balloon angioplasty (BA), and the article reviewed deals with the results of this procedure which should be carefully assessed. Both techniques, surgery and therapeutic catheterization, attempt to eradicate the effects of the disease via very different conceptual approaches. Surgery, through the total or partial resection of the stenotic segment or expansion of the area via grafting and BA, creates a "controlled rupture" of the vessel in the intimal and medial layers, with the aim of obtaining a favorable scarring process and later remodelling of the vessel. The efficacy of the treatment has been traditionally proved by the abolition of the pressure gradient between the upper and lower limbs and pressure normalization. All this is associated with the disappearance of the stenosis, demonstrated via angiography, magnetic resonance imaging, computerized tomography, etc.

This disease causes AHT and stimulates the formation of collateral circulation. Arterial hypertension seems to be basically related to the obstruction, although the possibility of associated and independent endothelial damage is not ruled out. However, the clinical presentation varies and depends on age and the frequently associated malformations. In the neonate or infant, it tends to appear with severe congestive heart failure. Interventricular communication, isthmus hypoplasia, aortic stenosis, bicuspid aortic valve, etc can also be present, adding risk, morbidity and mortality to the treatment. In the adolescent or adult, AHT is the predominant clinical feature that leads to diagnosis. However, AHT persists in 25%-40% of the cases treated, despite having eliminated the pressure gradient, and this will require pharmacological treatment for life.^{3,4}

Interventional cardiology, which is continually progressing, is attempting to replace the surgical solution with a "less aggressive" option, based on the potential dangers of surgery: mortality, paraplegia, effects of scar formation, pain, recoarctation, aneurysms and the overall economic impact, among others. Thus, its task is to demonstrate that this is feasible and with results comparable to surgical ones. However, it is important to bear in mind that surgery has already been used for

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60 years and that its long-term consequences are well known. The first BA was done by Singer in 1982 and the maximum follow-up time for this option is 23 years. Regarding treatment with stenting, the first implantation was done in 1991 and in 1995 Suárez de Lezo et al⁵ published the first large series. This means that the maximum follow-up time of this technique does not exceed 10-12 years.

From a merely anatomical standpoint, the surgical technique, with excision of the diseased wall, should be considered curative and is strongly advised, at least for young patients. This is not always possible and alternatives to end-to-end anastomosis are sometimes required, including patch aortoplasty. All surgical series have reported variable percentages of recoarctation and aneurysms, although the trend is decreasing. The Dacron patch seems to be associated with a greater incidence of aneurysms or late aortic dissections.

“Controlled rupture” of the aortic wall, which markedly weakens it, has been the method of treating this disease with BA. Initial series have already reported serious complications, ranging from restenosis, aneurysms and dissections, up to death, without forgetting “minor” flaps, residual gradients, vascular damage, and need for several redilatations or stenting at follow-up. In this issue of REVISTA ESPAÑOLA DE CARDIOLOGIA, del Cerro et al⁶ describe, in a group of native aortic coarctation patients with a mean age of 6.5 years, an immediate efficacy of 69% in the group of patients treated until 1988 and of 81.5% in the patients treated after this date. In recent series other authors have described remodelling in the treated vessel at follow-up, especially regarding localized coarctations, a lower frequency of aneurysms and a greater index of efficacy.^{7,8}

We have to admit that BA and stent implantation, initially or after previous angioplasty, are currently a favored approach and this is due to their apparent simplicity of use, speed and the small amount of material needed. Stenting avoids overdilatation of the aortic wall and leaves the pathological vessel wall in situ, with the hope that the scarring mechanisms will strengthen the vessel thus avoiding long-term problems.

Both techniques, although having good immediate results (BA) or very good results (*stenting*), need long follow-up before they can be compared to surgery. There is abundant medical literature on the treatment of aortic coarctation demonstrating that there is no single and definitive cure. The dilemma lies in choosing the most suitable method, in assessing when it should be applied and finding out how it could change the daily life of the patient.

The universally accepted indications for treatment are: systolic gradient at rest ≥ 30 mm Hg (>20 mm Hg under catheterization and sedation), AHT, impact on the left ventricle in the electrocardiogram or echocar-

diogram, and a pathological Holter blood pressure curve. Previous stroke or the presence of severe isthmic stenosis, as shown by angiographic imaging, nuclear magnetic resonance imaging, computerized tomography, etc, are also considered therapeutic indications. The indication for treatment should be unique for surgery or intervention, since both have a high risk of complications. In the first year of life surgical indication is undisputable and we currently have a series with excellent results: 181 infants operated on with just 4 recoarctations (2.2%), 3 of them with a weight lower than 2 kg.⁹ However, in this period BA is associated with a high incidence of restenosis (33% vs 26% in the study analyzed) and vascular damage is more frequent. The article by del Cerro et al⁶ shows that it is possible to successfully treat 85% of native coarctation patients but at the cost of serious complications (19.2%) in the first 26 patients and 3.7% in the last 27. Thus, it would be advisable to be able to establish, with valid criteria, which patients are good candidates for BA, and thus avoid using this technique in those patients at more risk of presenting complications.

Analysis of international results supports the value of BA in native coarctation, as an alternative to surgery, from the age of 1 year old until adolescence, but we should be aware that a percentage of patients will remain ineffectively treated. Furthermore, we still lack long-term follow-up in redilatated vessels. Balloon angioplasty, however, is considered the technique of choice regarding aortic recoarctation after surgery, due to its lower risk deriving from the greater perivascular scarring response.

From adolescence onwards, or in patients weighing more than 30 kg for some authors, the controversy focuses on 2 areas: surgery versus intervention, and BA versus primary implantation of stent. All the treatments have their supporters and detractors, showing that a standard treatment is still lacking. In addition, we know that the results can vary considerably depending on experience and that each center should treat patients with the technique they have most experience with. In the case of BA and stenting, given the potential complications of the procedure, it is essential to have anesthesia and thoracic surgery available in the center, as well as a complete stock of material (large-caliber sheaths, coated stents, balloons and stents of different lengths and diameters, rigid guidewires and arterial vascular sealants).

When analyzing our own experience, the article by del Cerro et al,⁶ and other medical literature, the strong impact of technological advances on the results can be appreciated, especially regarding BA. Thus, the relatively large balloons initially used were associated with the appearance of complications that occasionally were even fatal. In recent years, balloon profile has been considerably reduced (thus reducing the size of

the arterial introducer sheath), maximum inflation pressure increased, the passage of the balloon greatly improved, and they are now smoother with a shorter cuff. This fact is especially important in the pediatric population, where vascular complications are more frequent. We consider that the anatomy and the size of the aortic isthmus are essential to indicate, counter-indicate, or predict the effects of BA.¹⁰ In this regard, it is necessary to measure this segment very carefully (ideally in 2 planes, lateral and LAO 60°), and rule out dilatation in moderate or severe isthmic hypoplasia to avoid overdilating this area during inflation.¹¹ All these factors lead to improved results with this technique in the short- and medium-term. Finally, it is important to recall that BA is contraindicated in aortas with arteriopathy, as in Turner's and Marfan's syndromes, a fact also emphasized in the study by del Cerro et al.⁶ In these cases, the implantation of stents offers a good alternative to surgery.

Remarkable advances have been achieved with stenting. Since the initial descriptions by Suárez de Lezo et al⁵ and other investigators,^{6,12-15} the procedure has been standardized, with a reduction in the use of potentially traumatic and dangerous material (P-4014 and Palmaz series stents), which in some cases led to perforation of the aortic wall, with fatal consequences.¹³ Thus, the BIB (NuMed®) balloon and the CP stent¹⁴ are associated with greater efficacy and a reduction in complications. This stent has blunt edges, reasonable flexibility and wide cells, requiring 12 Fr introducers. Up to 2005, weld-fractures in the stent have been reported. The new prosthesis, strengthened with gold, is currently in use. Similarly, the Max LD stent from eV3® has wide cells which allow the introduction of sheaths of up to 12 Fr (personal experience), great flexibility and radial strength, but at the cost of an increase in the amount of metal which sometimes leads to some neointimal thickening.^{15,16} Stents with an expansion diameter less than the estimated size of the future aorta should not be used.

In recent years, maneuvers that offer a more aesthetic appearance (stent flaring or belling) have been advised against as they can adversely affect the end result.¹⁷ Some groups also recommend two-step stent deployment in very severe coarctations or the initial use of a coated stent in adults with acute coarctation. Unfortunately, the appearance of aneurysms has been demonstrated in all types of treatment, both interventionist and surgical.^{6,10,18,19} The size of these aneurysms could be defined as small: less than 3 mm; medium: <50% of the diaphragmatic aorta; and large: >50% of the diaphragmatic aorta.²⁰ In this regard it has been suggested that the different types of coated stents could provide a "definitive" solution.²⁰⁻²³ However, we have verified aneurysms after stent implantation²⁰ that required in-stent stenting to treat them.

The medical literature shows that the immediate results of treating this complex lesion are quite similar with both surgery and interventionism. Thus, mortality is lower than 1% with both approaches, recoarctation 0% to 15% postsurgery, 6% to 15% post-BA, and less than 5% with stenting. Aneurysms appear in 0%-5% post-surgery (up to 15% in old series with Dacron patch), in 4%-15% post-AB, and in less than 5% after stent implantation. Paraplegia has been described following both angioplasty and surgery, with a frequency of less than 0.5%.

Regarding follow-up, in a study of 235 adults with coarctation, whether treated or not, Oliver et al²³ demonstrated how the natural history of the aortic vessel can be complex. In 37 cases (16%) they found aortic wall complications: true ascending or descending aortic aneurysms, pseudoaneurysms, aortic rupture, dissection, fistulas, etc, even in the patients treated. In our experience,²⁰ with a young population with 12 years follow-up, the patients with bicuspid aortic valve and treated coarctation can develop aneurysmatic dilatation of the ascending aorta (1 out of 29 cases), requiring complex surgery during evolution. In addition, other patients, "suitably" treated with stents, have needed redilatation, a new coated stent due to fractures in the previous stent or due to aneurysms after treatment with coated stents.²⁰ Finally, we have also seen exceptional cases with 3 and 4 stents in the thoracic aorta, which has become a rigid tube as a consequence of difficulties during implantation.

The natural history of such patients undergoing interventionism is not free of debate. Taking into account the disorder described by Oliver et al²³ and the short follow-up time, we should assume that these patients will require indefinite control on a yearly basis with clinical check-ups, blood pressure control, etc, as well as sophisticated imaging studies, to rule out any possible recoarctation or silent vascular complications. In relation to this, we have to remind ourselves that imaging plays a key role in diagnosis, treatment strategy and follow-up, highlighting nuclear magnetic resonance imaging and computerized tomography with three-dimensional reconstruction, with systems with 16 or more detectors. Furthermore, aggressive antihypertensive treatment is very important as well as changing as much as possible the risk factors for coronary heart disease, which appear earlier in this group of patients.

In view of all this, despite the results described by del Cerro et al⁶ and other series, we still think that currently surgery is a valid and important method in the treatment of this disease, especially in the first year of life. Similarly the importance of BA in aortic recoarctation in adults with this disease and its treatment with stents, simple or coated, is already known. Indication for BA might be restricted to between 1 year old and adolescence and as an adjunct to surgery in the re-

maining patients. It is simple to do and offers acceptable short-term results; however, we do not know the future behavior of the treated vessel, which means that (in place of an optimal surgical intervention) it should be done with caution. In turn, stenting provides greater efficacy and better immediate results (in adults the success rate is higher than 90% and provides a greater decrease in pressure gradient), but this technique still has a short follow-up. Nevertheless, as mentioned at the beginning of this article, more than 25%-35% of the patients treated will continue to present high blood pressure levels and this should be treated to minimize the risk of complications due to late degeneration of the arterial wall. Stenting is linked to BA, and to possible immediate or late complications in this disease, with low morbidity and mortality. It has replaced primary surgery in some centers.

A refined technique and the use of new materials have led to a notable reduction in complications, both in BA and stenting. These results are valid for BA for up to 18 years, as shown in the article by del Cerro et al,⁶ although, the future evolution of this group of patients in the longer term is a question that future studies will have to answer.

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