Research Paper Medical Science

COARCTATION OF THE AORTA IN ELDERLY PATIENTS – SOLVED PROBLEM OR EQUATION WITH UNCERTAINTIES

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ABSTRACT

Coarctation of the aorta is a frequently observed congenital malformation that is normally diagnosed and treated in childhood. It is defined as aortic stenosis at the segment of insertion of ductus arteriosus. The therapeutic options are balloon dilation and/or stenting, or cardiac surgery. Currently existing data on the therapeutic approaches are to some extend still controversial. The results from short-term experience of aortic interventions look promising, especially in the case of elderly patients with high perioperative risk and those with unsuitable anatomy for intervention. It should also be noted that operation after stenting is a more difficult and risky approach. In the paper we discuss treatment guidelines, advantages and disadvantages of each therapeutic option.

KEYWORDS : aorta coarctation, cardiac surgery, interventional treatment

Coarctation of the aorta (CoA) is a common birth defect which is normally diagnosed and treated surgically in childhood. The incidence of malformations is 5-8% of all congenital defects. It is defined as a narrowing of the aorta, most frequently in the segment of insertion of ductus arteriosus. Diffuse (tubular) hypoplasia of a larger aortic segment, proximal to the left subclavian artery as well as obstruction in the abdominal aorta is more rarely observed. It is often observed in association with other cardiac and non-cardiac malformations, such as bicuspid aortic valve and ventricular septal defect, and some rare genetic heart syndromes. Uncorrected CoA in adults is registered in close relationship with manifestations of symptomatic systolic hypertension over the site of coarctation (upper body half) established by measuring the blood pressure of the brachial artery of both upper limbs and/or cardiac murmur. The morphological substrate of the pathological changes varies widely and includes both discrete stenosis distal to the left a. subclavia combined with vast collateralization, and hypoplasia of the aortic arch and istmus, commonly observed in newborns. In other cases the disease manifests itself as a prolonged tubular stenosis of the descending aorta – (thoracic segment). Total occlusion (interruption of the aortic arch with a complete lack of connection between the ascending and descending part of the thoracic aorta) is the worst case of coarctation. The incidence of this anomaly is 3 in 1 000 000 live births. If not corrected 90% of patients die in their first year of life. To date single instances of uncorrected total occlusion in adults has been described. Angiographically determined coarctation gradient ≥20 mm Hg defines hemodynamically significant CoA in the absence of well-developed collateralization.

The natural course of the uncorrected coarctation is premature death from stroke, coronary heart disease and sudden cardiac death, which prevention requires mandatory treatment (Campbell 1970).

Treatment of CoA

Therapeutic options are Interventional treatment or Cardiac surgery.

There is still no clear evidence of a unequivocal advantage of one over the other treatment option.

The aim of therapy is to perform a full restoration of conductivity of the aorta with minimal complications and gradual reduction of the residual gradient across the segment in the coarctation region.

Decision on the selection of a therapeutic approach is made depending on:

1. Age of the patient and severity of the pathology;
2. Anatomical and morphological features of the malformation;
3. Advantages and disadvantages of treatment methods;
4. Existing recommendations of specialized cardiology associations, European and American Heart Association (Table. 1 and 2)[1];
5. The experience and capabilities of relevant clinical centers.

There have been significant advances in the surgical treatment of CoA in the last 50 years. In the largest study to date of a series of cases on 646 operations, the perioperative mortality was the highest in infants, followed by children (aged 1-14 years), with a gradual increase in younger age and followed by a peak of around 4.5% in patients >30 years. Data on mortality and morbidity in elderly patients are significantly more scarce compared to the data for pediatric patients but suggest a higher procedural risk related to the technical parameters of the surgery and comorbidity, characteristic for older patients (Aris et al. 1999; Bouchart et al. 2000).

There are various cardiac surgical techniques depending on the length of the narrowing:

1. Small length: the narrowed segment is excised and aortic anastomosis is performed;
2. Average length: aortoplasty is performed;
3. Large length: bypass of the narrowed segment is performed.

Indications for performing cardiac surgery are: presence of chronic heart failure; uncontrolled systolic hypertension; peak gradient over the coarctation over 20 mmHg, measured by echocardiography or invasive unsuccessful intervention of CoA.

In women of childbearing age with native coarctation there is a problem with the integrity of the tissues in the paracoarctation region, especially in view of potential pregnancy. In these cases, surgery with excision of the paracoarctation tissue is preferred. Early mortality is less than 1% for primary surgery. Mortality after reoperation is higher (1-3%) and may reach 5-10% in comorbidity and heart failure.

Among the most significant complications of surgical treatment in adult patients with CoA are paraplegia in patients with inadequate collateral circulation (long cross-clamping) and damage to the spinal arteries (0.5-1%) (requiring full or partial cardiopulmonary bypass) (Conolly et al. 1998). Other complications include: paradoxical hypertension, postcoarctectomy syndrome, consequence of the organ reperfusion response, damage to laryngeal/phrenic nerve (6%), pleural effusion or thoracotomy complications. Re-coarctation (6%) and formation of aneurysms are also important, but late and less frequent complications.

Secondary prevention

Prophylaxis of infective endocarditis is performed up to 6 months after surgical correction of the aorta.

Interventional treatment

The development of interventional techniques led to an increase in the number of successful interventions in patients with CoA. In this regard, balloon angioplasty is widely used in the last 20 years. (Ovaert et al. 1998). The method is traumatic in its essence since there is an increase in the diameter of the vessel with interruption of the aortic intima and media, and development of cystic medionecrosis (Sohn et al. 1994). The degree of counter-resistance and recoarctation in response to balloon dilation results in a higher incidence of reinterventions, especially in the more expressed forms.

In earlier studies it was established that balloon dilation can lead to overdilation, which explains the observed higher incidence of formation of aneurysms - 5-20%, and rarely to fatal events of aortic rupture (Ovaert et al. 1998). A number of authors define patients with arch or istmus hypoplasia and tubular coarctation as unsuitable for angioplasty. For recoarctations after surgical correction opinions prevails in favor of dilation or stenting in the absence of aneurysm, pseudoaneurysm or significant coarctation, affecting arteries leaving the aortic arch.

According to data from different authors the success of intervention after angioplasty is defined as a gradient <20 mm Hg (Zabal et al. 2003). Cardiovascular risk, however, can increase also in moderate coarctation with no indications for intervention, but there is no data that gradient reduction below a certain threshold correlates with a relevant clinical benefit for the patient.

In connection with technology improvements there is also an increase in cases of endovascular stenting in patients with CoA (Harrison et al. 2001; Duke et al. 2001). In contrast to balloon dilation, the stenting method is based on the elastic properties of the aortic wall and is proven to reduce the degree of established recoarctation using only balloon angioplasty. The stenting method of aortic coarctation has limited opportunities for children in connection with the increasing diameter of the aorta, in parallel with increasing age and need for redilation, unlike its use in young unoperated adults and older children, where the method is a successful alternative to operational surgery.

It was established that stenting leads to lower residual gradient compared to angioplasty and lower incidence of residual hypertension during monitoring. There are other circumstances in favor of primary stenting in patients who have completed somatic growth. Instrumental aortic intervention after unsuccessful balloon dilation can provoke extensive dissection and increases the risk of subsequent stent disposition. Stents can reduce traumatic damage of the vascular wall by distributing the compression over a larger area, controlling the possibility of small dissections and formation of aneurysms.

Unresolved CoA stenting problems

There are many issues connected with the CoA stenting method regarding mainly the limiting factors of the procedure and long-term results:

- Use of coated stents - possibility of formation of aneurysms and dissections is reduced;

- Staged dilation – the healing effect between procedures is used in patients with unfinished somatic growth. The results from experimental animal models indicate a higher incidence of ruptures associated with the method;

- Application of rigid stents: further data regarding the use of rigid stents on cardiovascular hemodynamics is needed.

- The use of larger balloons in order to improve the positioning of the stent and epithelization of the aortic wall. Paradoxically, this practice may increase the risk of dissection due to the forced contact with the aortic media (Di Giovanni 2001).

Complications of interventional therapy

Despite aortic wall trauma, complications are rare (about 12%) and include:

Emergency surgery mortality after unsuccessful intervention is less than 1%. The rate of recoarctation and suboptimal postprocedural results after balloon angioplasty is approximately 7% in each case. The rate of late aneurysms is 4%, although the true incidence is unknown and depends on the diligence and monitoring possibilities of the patient. Other complications include: balloon rupture, incorrect stent positioning (4%), vascular complications in femoral access (3%), aortic dissection, myocardial infarction and stroke (1%). Most of these complications are minimized in the presence of an experienced team and a center for conducting the intervention, introduction of innovative stents, balloons and guides. Most severe complications require preparedness for cardiac surgery intervention with corresponding center proximity and staff availability.

Patient monitoring

The sensitivity of radiography and echocardiography for diagnosing aneurysms is below 100%, and that of echocardiography is further reduced by the presence of a metal stent. A routine contrast-enhanced spiral CT scan is recommended on the sixth week and the first year after stenting. An alternative monitoring method is magnetic resonance angiography. In patients with CoA there is an increased risk of developing ischemic heart disease, particularly in those with acquired CoA.

Undoubtedly there is an established postprocedural significant decrease in blood pressure (Marshall et al. 200). In experimental animals there were no observed side effects in normal aorta stenting. Despite the good results from experimental laboratory studies, it is unlikely that stenting will neutralize the reduced arterial compliance in the upper half of the body in patients with CoA, as well as the fact that the rigid stent can change significantly and adversely the vascular hemodynamics during physical load. Attention also requires the overall shorter monitoring period for evaluating the effect of stenting on the aortic wall. It has been shown that the loss of normal pulsatile movement in stenting alters the phenotype of the vascular smooth muscle cells and connective tissue by increasing the processes of apoptosis and subsequent formation of aneurysms (Pihkala et al 2001; Collum et al. 1997).

Practical guidelines

In adults with discrete CoA usually percutaneous balloon angioplasty is applied with or without stent implantation. The therapeutic success is 98%.

In conclusion, short-term experience and the results of interventional treatment of patients with CoA are promising, especially in cases of adults with high surgical risk. In native coarctation with suitable anatomy stenting is recommended as a method of first choice, but only if there is a center with adequate preparation. Cardiac surgery is an excellent option for the treatment of severe complex coarctation with istmus and arch hypoplasia as well as long tubular stenosis. In cases of failure of interventional therapy the use of extra-anatomic coronary aortic bypass should be considered. Operational therapy requires strict risk assessment in adults. Surgery after unsuccessful stenting is more difficult and risky. Surgical therapy is still the method of choice, having an advantage to interventional treatment of patients with CoA, despite existing recommendations and evidence of favorable results in stented and dilated patients.

Table 1. CoA treatment guidelines - European Society of Cardiology (ESC, 2014)

Interventional treatment	Class/ Level of evidence	Cardiac surgery

non-invasive pressure difference >20 mm Hg between upper and lower limbs, regardless of symptoms but with upper limb hypertension (>140/90 mm Hg in adults), abnormal blood pressure response during exercise, or significant left ventricular hypertrophy	I C	Inadequate anatomy (long and rugose/ convoluted segment)
Independent of the pressure gradient, hypertensive patients with >50% aortic narrowing relative to the aortic diameter at the diaphragm level (on MRI, CT or invasive angiography).	IIa C	
Regardless of the gradient and in the presence of hypertension in >50% narrowing of the aortic lumen in the absence of hypertension.	IIb C	

Table 2. AHA CoA treatment guidelines in elderly patients (2008)

Interventional treatment	Class/ Level of evidence	Cardiac surgery	Class/ Level of evidence
Peak-to-peak coarctation gradient greater than or equal to 20 mm Hg	IC	Unsuccessful intervention	IB
Peak-to-peak coarctation gradient less than 20 mm Hg in the presence of anatomic imaging evidence of significant coarctation with radiological evidence of significant collateral flow	IC	Long recoarctation segment	IB
Recoarctation	IB	Concomitant hypoplasia of the aortic arch	IB
Stent placement for long-segment coarctation may be considered, but the usefulness is not well established, and the long-term efficacy and safety are unknown.	IIbC		

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(Endnotes)

1 Existing guidelines do not discuss treatment in elderly patients with total 6occlusion.