



Treatment for coarctation of the aorta: where are we and where do we go from here?

“Even the most advanced technology, such as growth stents and absorbable stents, cannot treat the systemic vasculopathy as such, but the anatomical treatment facilitates adequate blood pressure treatment.”

KEYWORDS: arteriopathy ■ balloon angioplasty ■ coarctation of the aorta ■ pharmacological treatment ■ stenting ■ surgical treatment

Coarctation of the aorta

Coarctation of the aorta is typically a stenosis of the descending aorta in the area of the ductus arteriosus Botalli and caused by ductal tissue in the aortic wall. Once the duct constricts post-natally, this tissue constricts as well and leads to the typical posterior shelf, which is visible on echocardiography. Aortic coarctation is found in approximately 1:3000 newborns and, if left untreated, reduces the life expectancy to 34 years on average [1]. Associated congenital heart disease such as bicuspid aortic valves, hypoplastic aortic arch, ventricular septal defects and others are frequently observed [2]. Clinical signs are weak or absent femoral pulses and blood pressure difference from upper to lower limbs. A heart murmur may be audible on the left sternal border, but it is more important to listen between the scapula. While in neonates echocardiography alone is sufficient to establish the diagnosis, as the treatment is typically surgical, later in life further imaging such as MRI or computed tomography scans will allow adequate therapeutical planning.

Surgical treatment

Since 1944, coarctation of the aorta has been treated surgically [3], and nowadays the most commonly used method is the end-to-side anastomosis, during which the coarcted segment is cut out and the ends are joined. A modification is the end-to-side anastomosis. Further surgical techniques have been applied such as patch plasty, direct plasty, tube grafts, subclavian flap or an extra anatomic bypass. The life expectancy is overall better and long-term survival can be achieved in up to 80% of cases [4].

Interestingly, a re-stenosis occurs in 15–20% of all cases independent from the type of surgery, although some centers report lower rates [5]. Aneurysm formation is relatively rare, but has

the potential of rupture and should therefore be treated aggressively. It is noteworthy that 50% of all patients after coarctation surgery in the long term require antihypertensive medication. The hypertension may be subclinical (no resting gradient on echo but on exercise an increased gradient) [6].

Angioplasty

As the results are suboptimum with regards to the re-coarctation and rate of hypertension, it is no wonder that the pediatric cardiac interventionalists thought of a different approach and invented angioplasty in the early 1980s [7]. The results here were quite dependent on the age at intervention: if it was performed in neonates, the re-coarctation rate was quite high (30% and above) [8]. If it is performed after the first year of life, the re-coarctation rate is comparable to surgery. Aneurysm formation seems to occur a bit more frequently than in surgery. This can occur both in the operative or in the interventional field as well, proximally or distally to the coarctation area [7].

Stent implantation

To avoid a recoarctation due to re-coil, stents were introduced into the treatment armamentarium for coarctation in the early 1990s. Nowadays several different types of stents with different strut design (closed cells vs open cells) and bare-metal stents and covered stents are used for treatment of the coarctation. Even very severe and sub-atretic coarctations can be treated with stents nowadays [9].

After angiography to measure the vessel size at the transverse arch and at the level of the diaphragm as well as in the coarctation area, a guide wire is positioned either in the left ventricle or left subclavian artery. The femoral sheath is then



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exchanged to a long delivery sheath, through which the stent–balloon ensemble is forwarded into position. Once good position is established with multiple hand angiographies, the balloon is inflated, and with this the diameter of the coarcted segment is increased and the stent is pressed into the vessel wall to achieve secure positioning. Complications can occur as well, the majority of them are access related at the femoral or inguinal arteries. Stent fractures have been described and aneurysm formation can occur at the site of a bare metal stent, or proximally or distally to the coarctation site.

“Interventional treatment, as well as surgical treatment, can only deal with the anatomical coarctation.”

An ongoing problem in all interventions is the arterial hypertony, which warrants medical treatment in many of the cases. In our own recently published series of 68 patients with long-term follow-up of up to 13 years, we could show that after stent implantation, only a third of patients needed antihypertensive medication, but during follow-up this increased to approximately 50%. After re-dilation of the stents, which was carried out on 26 patients, there was only need for medication in 35%, but in the long-term follow-up, two thirds of the patient needed antihypertensive medication [10]. This is comparable to other data from the literature.

Systemic vasculopathy

The high rate of hypertension independent from the type of treatment led to the concept of a systemic vasculopathy. Indeed, even in neonates it could be shown that the aortic distensibility is impaired and the stiffness is increased at least in the proximal vessels. This seems to be progressive over time [11,12]. It could be shown that 35 years after coarctation surgery the flow induced dilatation of the aorta and the nitroglycerine

induced dilatation is decreased, while the pulse wave velocity along the vessel wall is increased (indicating a stiffer vessel wall) [13].

Several markers have been tested to see whether there is endothelial dysfunction. It seems that the soluble intercellular adhesion molecule one and the IL-10 are raised after coarctation repair, which led to the conclusion that there is late inflammatory reaction [14]. The asymmetric dimethyl arginine, as an endogenous competitor of the nitric oxide synthetase level, is raised after coarctation repair, which is a marker of vascular dysfunction and, as less endogenous nitric oxide is produced, the distensibility of the endothelial may be impaired. It is also known that in conditions where this marker is raised, there is a higher risk for coronary and cerebral pathology [15].

It seems that after coarctation repair, aggressive follow-up, including blood pressure measurements and medical treatment for arterial hypertension is necessary. Therefore, especially in the pediatric population knowledge of normal values for age and size is paramount [16].

Interventional treatment, as well as surgical treatment, can only deal with the anatomical coarctation. Even the most advanced technology, such as growth stents and absorbable stents, cannot treat the systemic vasculopathy as such, but the anatomical treatment facilitates adequate blood pressure treatment. Prior to intervention any antihypertensive medication may lead to a hypotension in the post stenotic area, but after any type of anatomical correction achieving and maintaining normotension in order to avoid long term consequences of hypertension seems paramount. Looking at the arteriopathy, it seems that angiotensin converting enzyme inhibitors should probably be used as first line medication and not β -blockers. Further research is definitively necessary to enhance our so far incomplete understanding of the ‘simple’ congenital heart disease coarctation of the aorta.

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