Original research article

Endovascular management of coarctation of the aorta in adult patients

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A R T I C L E   I N F O

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A B S T R A C T

Background: Coarctation is a congenital heart defect with an incidence of 5–8%, more frequent in boys; however, the overwhelming majority of patients undergo surgery earlier as pediatric patients. Currently, the treatment of choice in adolescents and adults is endovascular management.

Method: We used all three common catheter-based techniques: simple balloon dilatation, stent placement, and stent-graft placement. All treated patients underwent pre- and post-procedural CT angiography.

Group of patients: Between 2004 and 2014, we treated a total of 10 patients with coarctation. They included seven men and three women, with a mean age of 41.1 (21–59) years. Eight patients were treated for native coarctation, and two for re-coarctation. Dominant features included hypertension (eight patients) and left-sided heart failure (four patients); some patients presented with multiple conditions at a time. Simple dilatation was performed in two patients while four had both stent and stent-graft placement.

Results: All procedures were technically successful, with a long-term beneficial effect seen in all patients. All patients with hypertension showed improvement, with medication either reduced or completely discontinued in four cases.

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Introduction

The term, coarctation, refers to a narrowing of the aortic isthmus or, in rare cases, of the thoracic or abdominal aorta. It is a local ridge bulging from the posterior and lateral aortic walls toward the ductal orifice, and formed by thickened media or, possibly, intima. Coarctation occurs in 5 to 8% of congenital heart defects, more commonly in boys than in girls, at a ratio of 2:5:1 [1–3]. Its exact cause is unknown. Coarctation should be distinguished from tubular hypoplasia, the latter being a narrowing of the proximal segment of the aortic arch to 60%, distal aorta to 50%, and aortic isthmus to 40%. Unlike coarctation, tubular hypoplasia is characterized by a normal wall structure. Critical aortic narrowing in infants and neonates is associated with completely different hemodynamic and clinical presentations, compared with coarctation in older children and adolescents. Neonates require urgent treatment, which is predominantly surgical given the long-term

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failure rate of balloon dilatation [4]. The currently preferred technique used in adolescents and adults is endovascular treatment, particularly in those with re-coarctation. Our paper presents our experience with this technique.

Materials and methods

All our patients underwent pre-procedural CT angiography and anatomical findings were used to decide on the size of material and technique employed. All patients underwent scheduled, non-urgent procedures.

We used the three standard techniques for catheter-based management: simple balloon dilatation, balloon dilatation with stent placement, and stent-graft placement with additional balloon dilatation. In each case, the decision on the particular technique to be used was made based on the following criteria: (a) simple dilatation with stenoses of borderline significance, anatomically unsuitable for stent placement; (b) stent placement with significant stenoses localized close to arch branches, and in younger patients; (c) stent-graft placement with tight stenoses, older patients, and post-stenotic dilatation. All procedures for endovascular management began with catheterization, an uncomplicated task with less significant stenosis. A challenging problem was trying to pass through the very tight stenoses and post-stenotic dilatation. In this case, we had to use a pre-shaped catheter and to manoeuvre a steerable guide-wire within the dilated aorta, which can be difficult, given the small slit provides poor support for the catheter.

The therapeutic strategy changed over the 10-year period, with more sophisticated instruments being used. While simple dilatation was commonly used in the early endovascular era, the currently preferred technique is stent or stent-graft implantation. As the original stents required the use of thick 18–20 F sheaths, actual placement was performed from the femoral artery – similar to stent grafts – using a cut-down approach. Previously, we used custom-made, steel, self-expandable stents made by ELLA, a Czech manufacturer based in Hradec Královo. At present, we use nitinol, laser-cut, self-expandable Sinus-XL stents (OptiMed, Ettingen, Germany) adapted for 0.035-inch sheaths. The stent size ranged from 28 to 36 mm in diameter, and 40 to 60 mm in length; given the availability of 10 F sheaths, there was no need for groin cut-down. The procedures were performed exclusively under local anesthesia, with bleeding after sheath removal stopped by compression using a Femostop system (OptiMed, Ettingen, Germany). The same approach was adopted with balloon dilatation using 7 F sheaths for balloon catheters measuring 14 mm in diameter. The type of anesthesia used with stent grafts and stents requiring inguinal cut-down was at the anesthesiologist’s discretion, depending on whether epidural anesthesia or general anesthesia was performed. Given the pain associated with dilatation, particularly with tight native stenoses, patients received analgesics (opiates) during the procedure, after which administration continued for as many as several days post-procedurally. Heparin was only administered in patients who developed pelvic artery obstruction that resulted in blood flow reduction due to the thick sheath systems. The stent grafts used in our center were either Valiant (Medtronic, Santa Rosa, CA, USA) or Relay (Bolton Medical, Inc., Sunrise, FL, USA), invariably using the shortest lengths available (100 mm), as all the lesions were short. Both products are identical in design (endograft) featuring a nitinol skeleton on a woven fabric (Dacron). All stent grafts were tubular and of the same width at their proximal and distal ends. The stent-graft diameter was invariably chosen in order to be 20% larger than the diameter of the aorta at its anchoring site.

When opting for simple balloon dilatation, the technique was similar to that employed in percutaneous transluminal angioplasty. Caution had to be exercised in keeping the necessary dilatation pressure in the balloon, since the large-diameter balloon catheters are more prone to rupture. Even tight stenoses were successfully dilated at pressures of 4–6 atm.

When using a stent, dilatation was first undertaken with a smaller-diameter (usually 10–12 mm) balloon, followed by stent placement. The stent diameter was about 10% larger than that of the aorta. The dilatation was subsequently performed using a larger balloon, measuring up to 24 mm in diameter and invariably 20–30% smaller than the diameter of the intact aorta.

The third option for endovascular treatment was stent-graft placement via the femoral artery. Again, the first step was to dilate the tight aortic stenosis with a smaller (10–12 mm) balloon catheter, with the partly dilated stenosis subsequently covered with the fabric component of the stent graft. When necessary, the left subclavian artery was covered, either completely (in patients with a patent right vertebral artery and developed circle of Willis) or partly, to make sure brain perfusion would not be impaired. Prior to opening the stent graft, systolic blood pressure was decreased to a level below 100 mmHg to avoid possible stent-graft migration due to blood flow. Next, the stenosis site was dilated using a balloon catheter, again 20–30% smaller than the diameter of the intact aorta.

Prior to and after the procedure, we performed a direct measurement to determine the pressure gradient, firstly as a confirmation of indication of the procedure, and secondly as the main indicator of technically successful coarctation management. The minimum level of the systolic pressure gradient for the endovascular procedure was 20 mmHg. The measurement was most often made by withdrawal of a pigtail catheter with the values measured above and under stenosis. Less often, pressure was measured with the tip of the catheter above the stenosis in the aortic arch, with the second pressure value obtained from the sheath with the distal end in the iliac artery. Pre-procedurally, the pressure gradients ranged between 20 and 100 mmHg, with the highest post-procedural gradient being 12 mmHg. Our definition of technical success was absence of narrowing of the stent graft (non-significant at most) in the form of residual stenosis or recoil and decrease in the systolic pressure gradient at less than 15 mmHg.

Clinical success was defined as more readily controlled hypertension (reduction of antihypertensive medication or non-pharmacological normalization of blood pressure), absence of claudication or prolongation of the claudication interval, and remission of signs of left-sided heart failure. Post-procedural follow-up included CT assessment. The first
assessment was scheduled at least 3 months after the procedure, with subsequent examinations at a 1-year interval in first patients. After gaining some experience with the procedure and mainly due to the absence of any complications leading to new interventions, we changed our strategy to perform CT FU within the first 3 months only. The examination always included CT angiography with reconstructions assessing stenosis significance, and stent and stent-graft integrity. Also assessed were the size and number of collateral arteries. Further CT imaging would have been indicated only if there had been any clinical symptoms present, leading to suspicion of restenosis or other late complications.

Fig. 1 – (a) CT angiogram of the thoracic aorta with coarctation; (b) angiographic findings prior to dilatation; (c) balloon catheter-based dilatation; (d) follow-up angiography with a stent graft placed at the site of original coarctation; (e) follow-up CT 4 days post-procedurally with an insignificant residual stenosis; (f) another CTA assessment at 8 months – completely normalized findings.
Table 1 – Group of patients. Number of patients, lesion type, reason for management, and technique used.

<table>
<thead>
<tr>
<th>Patient M/F</th>
<th>Coarctacion</th>
<th>Re-coarctation</th>
<th>Hypertension</th>
<th>Cardiac failure</th>
<th>Pseudoaneurysm</th>
<th>Claudication</th>
<th>Balloon</th>
<th>Stent</th>
<th>Stent graft</th>
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<td>1</td>
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</table>

Patients

A total of 10 patients were treated in our center between 2004 and 2014. They included seven men and three women with a mean age of 41.1 (age range, 21–59) years. Eight patients had primary treatment of native coarctation, and two underwent re-coarctation treatment. All procedures were elective. All cases of native coarctation were incident findings, while, in the two cases of re-coarctation, the patients were lost to follow-up after surgery in childhood. Simple dilatation was performed in two patients, and four patients underwent both stent and stent-graft implantation (Fig. 1). An anesthesiologist was available for all procedures requiring groin cut-down, which applied for four stent-graft placements and two stent placements. Four procedures were undertaken under local anesthesia. Prior to endovascular treatment, all patients were assessed by a cardiologist, which revealed clinical symptoms of hemodynamically significant stenosis confirmed by CT angiography. The reasons for treatment are summarized in Table 1, the most frequent being hypertension in eight cases, followed by left-sided heart failure. One patient with re-coarctation and a postoperative pseudoaneurysm was found to have a small sac without mural thrombosis. Another patient experienced claudications after several hundred meters, which posed a major limitation in his case, as he was a young athlete.

Results

The procedure was technically successful in all our patients, with stents and stent grafts implanted in sites as necessary and dilatation balloons expanded as required. There were no fatal intra-procedural complications (Table 2) and no periprocedural deaths. A short, approximately 3 cm long dissection at the site of dilatation, distally from the stenosis, was noted in one case of simple balloon dilatation; the dissection was left untreated. The origin of the left subclavian artery was covered with a stent graft in two cases, neither of which required revascularization of the covered artery.

At present, the mean follow-up period is 4.1 years. All patients have had follow-up assessment by a cardiologist and at least one has undergone follow-up CT angiography. A long-term beneficial therapeutic effect has been seen in all 10 patients. There was one death 3 years post-procedurally not related to coarctation management. The procedure has never been repeated, nor has surgery been required. All eight hypertensive patients showed improvement, with reduced doses of antihypertensive drugs and antihypertensive therapy completely discontinued in four cases. One patient was treated with simple dilatation, while having significant re-coarctation documented on a follow-up CT angiogram, also showed clinical improvement and thus did not require aggressive management. The other CT-angio findings are summarized in Table 3. Claudications resolved in one patient and a pseudoaneurysm in another had been covered by a stent graft and are considered as treated. Follow-up examinations have not documented any changes in the short dissection, which developed during dilatation. Long-term findings in patients with stent or stent-graft placement remain unchanged, with no signs of impaired stent or stent-graft integrity.

Discussion

The therapeutic strategy in our center has evolved in parallel with the development of materials available for endovascular

Table 2 – Outcomes and complications of coarctation management.

<table>
<thead>
<tr>
<th>Technical success</th>
<th>10</th>
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<tbody>
<tr>
<td>Periprocedural death</td>
<td>0</td>
</tr>
<tr>
<td>Rupture/aneurysm/dissection</td>
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<tr>
<td>Embolism/vascular complications</td>
<td>0/0</td>
</tr>
<tr>
<td>Stroke</td>
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<tr>
<td>Groin complications</td>
<td>0</td>
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</tbody>
</table>

Table 3 – Follow-up CT after treatment.

| Restenoses | 1 |
| Significant | 2 |
| Insignificant | 7 |
| None | 0 |
| Wall/mural lesions | 0 |
| Aneurysm | 1 |
| Dissection | 1 |
management of the thoracic aorta region. The first technique we used was simple balloon dilatation, which, while considered safe and effective in the management of native coarctation of the aorta, is associated with a significant incidence of re-coarctation. Therefore, we decided to experiment by indicating a less tight stenosis for balloon angioplasty. Repeat dilatation was possible [4] and consequently, both patients managed by simple balloon dilatation did not show ideal findings on follow-up CT assessments, one developing a significant re-stenosis and the other, a short dissection. Although consensus has not been reached on whether or not this is an appropriate technique (surgery vs. balloon angioplasty) for native aortic coarctation, it is generally accepted that endovascular access is the method of choice for re-coarctation [4–7]. In the following period, the evolution of custom-made stents allowed us to extend our indications to a very tight stenosis. The first ever stent-based management of coarctation of the aorta was undertaken in early 1990. Stent placement helps decrease the incidence of acute elastic recoil and late re-stenosis [1]. Most recently, we have begun to employ stent grafts and commercially available stents. Use of stent grafts is recommended for the treatment of coarctation, in particular when combined with the following conditions: patent ductus arteriosus, Takayasu arteritis, very tight stenoses, aneurysms, marked tortuosity of the aorta, and cases associated with a high risk of aneurysm formation or development of dissection. In our center, stent grafts were used in patients with very tight stenoses. While we have not yet had a single patient with complete aortic atresia, there have been reports of coarctation resulting in a completely severed aorta, although producing a good outcome when using a stent graft [8]. As a preventive measure to manage potential rupture, it is our policy to always have a stent graft at hand even when we are planning to place a stent only. The technical success rate in all of our patients having stent or stent-graft implantation is comparable with data reported by other authors; the long-term beneficial effect is also consistently good. Unlike other reports, quite surprisingly, native coarctation was more frequent than re-coarctation in our patient group [9–12]. Overall, the patient sets published to date have been small, despite being reported by large center reports [13,14].

Use of modern stents with suture systems and low-profile sheaths to manage an unprepared groin when using a stent graft allows the procedure to be performed under local anesthesia, where the role of the anesthesiologist is confined to analgesedation. An issue yet to be addressed is further management of patients, particularly the insertion of a central venous catheter (mostly not used). Central venous access is critical in order to make up for blood loss management, should a rupture occur. While rare, aortic rupture is a fatal complication; however, only four cases have been reported to date [15]. As anesthesiologists are able to better relieve chest tenderness associated with dilatation, we consider their presence a definite advantage. While the risk of intra-procedural complications is substantially reduced by detailed knowledge of surgical procedures performed in childhood, the unavailability of patients’ medical records poses a major problem.

No clear-cut recommendations regarding follow-up of patients with coarctation, especially follow-up using imaging techniques, have yet been published. Computer-tomography-based follow-up at a 5-year interval has been suggested [16,17]. Computer tomography-based follow-up of most of our patients with stent or stent-graft implantation showed either complete normalization of the aortic diameter or only an insignificant residual stenosis, with diminished collateral circulation (Fig. 2).

![Fig. 2](image-url)
Long-term follow-up of our first patients with stent placement has revealed no major changes. None of the patients have experienced undesirable events, such as rupture or re-coarctation. Although given the risk of dissection this subgroup of patients should be preferably followed up by CT in the early postprocedural period only, CT-based follow-up will not provide any further new information. Based on the radiation exposure in young patients on long-term follow-up, it is our recommendation to perform CT assessment only in cases where there is a change in the clinical findings. Patients undergoing simple balloon dilatation should preferably have annual CT follow-up given their higher risk of a restenosis, aneurysm formation, or dissection [1].

**Conclusion**

As also confirmed by our results, endovascular treatment of coarctation is a safe and rapid technique posing only a little burden to the patient. Another advantage is the short hospitalization time. In our center, surgical treatment is reserved only for cases of complex anatomies that are not suitable for catheterization.

**Conflict of interest**

There is no known conflict of interest associated with this publication.

**Ethical statement**

This publication has been prepared according to the ethical standards approved by our Institute’s Ethical Committee.

**Informed consent**

All patients and volunteers who participated in this research gave their consent for the results and images to be published.

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**References**