Institutional review - Congenital

Extra-anatomic aortic bypass for complex (re-) coarctation and hypoplastic aortic arch in adolescents and adults

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Abstract

Various surgical approaches have been proposed for complex (re-) coarctation and aortic arch hypoplasia (AAH). We report seven patients (mean age 19.6 ± 9.5 years) with complex coarctation or re-coarctation and AAH successfully treated by extra-anatomic ascending-to-descending aortic bypass (ADB) via sternotomy between 1995 and 2002 without mortality and no relevant complication early postoperatively and during a follow-up of 24 ± 29 (0.2–84) months. ADB may therefore be considered in selected patients with (re-) coarctation with AAH, with the need for concomitant ascending aortic or cardiac surgery and in patients with aortic arch stenosis and increased risk of complications under DHCA.

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1. Introduction

Recurrent coarctation may be difficult to manage. If associated with calcification and/or diffuse tubular hypoplasia of the aortic arch, percutaneous balloon dilatation and stenting will not be successful. Most surgical options, including resection with direct end-to-end anastomosis, patch aortoplasty, or interposition tube grafts, require cardiopulmonary bypass (CPB), exposure of the previously operated aortic segments and aortic clamping with inherent risks of bleeding, injury of adjacent nerves, and paraplegia. In cases of concomitant hypoplastic aortic arch, deep hypothermic circulatory arrest is needed for augmentation of the stenotic segment, exposing the patient to increased risk of cerebral dysfunction, cerebrovascular accident, and increased bleeding. Extra-anatomic ascending-to-descending aortic bypass however, does not need local dissection of the coarctation or aortic arch, clamping or CPB and yields excellent short- and mid-term results.

2. Material and methods

Seven patients operated on between 1995 and 2002 with an ascending–descending aortic bypass (ADB) graft for complex coarctation and aortic arch hypoplasia (AAH) were retrospectively analyzed. Patients’ characteristics are summarized in Table 1. Preoperative assessment usually comprised echocardiography, Angio-computed tomography (CT) or angio-magnetic resonance imaging (MRI) (Fig. 1). Invasive aortography was performed in two patients with aortic balloon dilatation and stent placement in both.

2.1. Surgical technique

Surgery is performed via sternotomy and the descending aorta exposed retropericardially by gentle luxation of the heart, using techniques known from off-pump CABG surgery. After systemic heparinization, the descending aorta is partially side-clamped and an end-to-side anastomosis with a ring-reinforced ePTFE graft (ImpraFlex, Impra Inc., Tempe, AZ, USA) or a Vasutek Gelweave graft (Vasutek Ltd., Renfrewshire, UK) performed. After local control for hemostasis, the posterior pericardium is re-adapted. The graft is positioned along the inferior aspect of the heart and the right atrium to the ascending aorta. In growing patients, additional length of the graft may be gained by opening the pleural cavity to avoid growth-induced tension. After side-clamping of the ascending aorta, the proximal end-to-side anastomosis is performed, the graft de-
aired and de-clamped. The operation is terminated with closure of the pericardium and the chest in a standard fashion. All patients are routinely followed in our outpatient clinic and periodically assessed with CT scan or MRI (see Fig. 2). Anti-hypertensive medication is usually continued for 3–6 weeks postoperatively and then discontinued in normotensive patients.

3. Results

Seven patients with a mean age of 19.6 ± 9.5 (11–38) years were operated on between 1995 and 2002. All but two patients suffered from re-coarctation and concomitant severe AAH and had undergone multiple previous corrective procedures. All except one patient had upper body hypertension and two suffered from leg claudication. One patient (case 2) developed a pseudoaneurysm at the distal anastomotic site with hemoptoe after anteriorly placed ADB 7 years before. One patient (case 6) was managed with a hybrid procedure of ascending–descending bypass, transposition of the left common carotid and subclavian artery to the ascending aorta and interventional stenting of an isthmic aneurysm. In two patients placement of a ADB was their first surgical intervention: In one female patient presenting with a severely hypoplastic distal arch and a long coarctation, CPB and DHCA was avoided because she suffered from phacomatosis with av-malformations of the cerebral vessels and internal carotid aneurysm. In the other patient (case 7), concomitant replacement of an ascending aortic aneurysm and placement of a ADB to treat severe hypoplasia of the mid and distal arch with coarctation was performed via sternotomy.

Mean operation time was 217 ± 136 (120–510) min and ICU stay 1.4 ± 0.5 (1–2) days. Mortality was 0% in this small series and the postoperative course was uneventful in all patients. They were discharged home after a mean length of stay of 10.4 ± 4.1 (6–16) days. During a follow-up period of actually 24 ± 29 (0.2–84) months, no adverse event was noticed and anti-hypertensive medication was stopped in all but the one patient, who was recently operated on. Peri- and postoperative data are presented in Table 2.

4. Discussion

The prevalence of recurrent coarctation varies widely from 7 to 60% [1] of operated coarctations and may be difficult to manage. In this situation, the procedure of choice is percutaneous balloon dilatation and stenting [2]; however, with calcification and/or diffuse tubular hypoplasia of the aortic arch this approach may not be recommended. Various surgical options have been proposed to deal with recoarctation including resection of the narrowed segment with direct end-to-end anastomosis, patch aortoplasty, interposition tube grafts, bypass grafts from the aortic arch or subclavian artery to the descending aorta. Most of these procedures require cardiopulmonary bypass (CPB) in the adult and dissection of adhesions with an inherent risk of bleeding and injury of adjacent recurrens and phrenic nerves, while cross-clamping of the aorta near the narrowed segment may be hazardous, since the aortic wall is abnormal [3] and frequently calcified.

Table 1
Patient characteristicsa

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Diagnosis and indication</th>
<th>Grad</th>
<th>Previous procedures</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>38</td>
<td>M</td>
<td>Recoarctation (stenotic graft segment), diffuse AAH; upper body hypertension</td>
<td>50</td>
<td>Coarctation repair (Dacron tube graft interposition) at 16y; Dilation and stent</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(Wallstent 16 × 37 mm) at 36y; Coarctation repair (Dacron patch plasty) at 7y;</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>re-coarctation and AAH treated with ADB at 15y</td>
</tr>
<tr>
<td>2</td>
<td>22</td>
<td>F</td>
<td>Large anastomotic pseudoaneurysm descending aorta; hemoptoe</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>12</td>
<td>F</td>
<td>Severe AAH with distal occlusion, severely hypoplastic descending aorta; upper body hypertension</td>
<td>120</td>
<td>Coarctation repair (ete) at 1y</td>
</tr>
<tr>
<td>4</td>
<td>14</td>
<td>M</td>
<td>Distal AAH; upper body hypertension</td>
<td>45</td>
<td>Coarctation repair (ete) at 5y</td>
</tr>
<tr>
<td>5</td>
<td>11</td>
<td>F</td>
<td>Phacomatosis with atrio-venous malformations of brain vessels and lower extremities, left internal carotid artery aneurysm, severe distal AAH and long coarctation segment; upper body hypertension</td>
<td>50</td>
<td>–</td>
</tr>
<tr>
<td>6</td>
<td>21</td>
<td>F</td>
<td>Severe hypoplasia of the mid and distal aortic arch, aneurysm at the coarctation repair site; upper body hypertension</td>
<td>95</td>
<td>Coarctation repair (pericardial patch plasty) at 3w, restenosis redo at 8y, dilation and stent (Wallstent) of AAH, aneurysm at 19y</td>
</tr>
<tr>
<td>7</td>
<td>18</td>
<td>F</td>
<td>Severe mid and distal AAH, severe coarctation, aneurysm of the ascending aorta; upper body hypertension</td>
<td>100</td>
<td>–</td>
</tr>
</tbody>
</table>

aAAH, aortic arch hypoplasia; ADB, ascending–descending aortic bypass; ete, end-to-end anastomosis; Grad, systolic gradient; y, years; w, weeks.
Furthermore, the patient is exposed to the risk of perioperative paraplegia, which should not exceed 3–5%. Extra-anatomic ascending-to-descending aortic bypass, however, does not need local dissection nor cross-clamping of the diseased aortic segment, nor CPB, and yields excellent short- and mid-term results in published series [4–9]. Some patients with so-called re-coarctation have rather AAH than restenosis at the original coarctation site. This might be due to inadequate initial repair with failure to address the problem of arch hypoplasia present in 30% of coarctation [10,11] (Amato type II coarctation) or inappropriate growth of the aortic arch. Not surprisingly, AAH has also been identified as risk factor for re-coarctation [12]. Surgical correction may be achieved by local patch enlargement with the use of deep hypothermic circulatory arrest (DHCA), exposing the patient to its inherent risks of cerebral dysfunction, cerebrovascular accident, coagulation disorders and bleeding. These risks may be increased in certain patients, like in our patient with av-malformations of the brain vessels (case 5). Surgery is usually performed via sternotomy and distal exposure of the aorta is limited to the isthmus region. Should a concomitant especially long re-coarctation be treated as well, one may not be able to carry the distal extension of the patch far enough to adequately enlarge the coarctation, nor would it be possible sufficiently mobilize the descending aorta should a local resection and end-to-end anastomosis be envisaged. In this case, exposure through a left re-thoracotomy may be considered, however again with the need for CPB with DHCA, local dissection of adhesions and the risk of injury to adjacent anatomical structures or the diseased aorta. With the technique of ADB DHCA and its risks, as well as the need for local dissection of a previously exposed area, are altogether avoided.

Mortality rates reported in the literature for direct repair of re-coarctation vary between 0 and 7% [12,13]. Thus, the 0% mortality rate of the present and other series [6,9,14,15] compares favorably, although others report mortality rates of 7–17.7% [7] for the ascending–descending bypass technique.

We favor placing the graft along the right atrium and the inferior aspect of the heart to the descending aorta. In children, additional length of the graft can be gained by opening the pleural cavity to avoid growth-induced tension. However, the bypass graft may alternatively be placed along

Fig. 1. Preoperative MR angiography showing recoarctation of the isthmic aortic segment (asterisk) and diffuse stenosis of the distal aortic arch (arrows).

Fig. 2. Postoperative MR angiography showing an excellent result after insertion of a long ascending–descending aortic bypass graft (arrows).
the aorta, which may be advantageous in case of future heart surgery.

One patient (case 2) developed an anastomotic pseudoaneurysm 7 years after anterior ascending—descending graft at the age of 15. This long-term complication, arising in up to 6% in published series [6,9], may have been favored by growth-induced tension on the anteriorly placed graft and the fact, that the distal anastomosis had been made at the site of the original coarctation. To avoid diseased proximal aortic segments the anastomosis should therefore be placed as far distal as possible on the descending aorta, exposure of which is facilitated by techniques used in off-pump CABG surgery to luxate the heart. Even in the patient with occluded proximal and extremely hypoplastic distal descending aorta (case 3), the anastomosis of the graft could easily be made above the diaphragm. In no case was opening of the peritoneum or exposure of the suprarenal abdominal aorta necessary to reach a graftable aortic segment.

Individual tailoring of the surgical approach is essential to reach good results and the technique of ascending—descending bypass is one of several that may be considered in an individual patient. In younger children, we would always try to anatomically reconstruct the aortic arch and isthmus, if technically feasible. We may favor the ADB in adolescent and adult patients with re-coarctation and aortic arch hypoplasia, coarctation or re-coarctation with arch hypoplasia and the need for concomitant ascending aortic or cardiac surgery and in patients with aortic arch stenosis and increased risk of cerebrovascular accident under DHCA. It may also be a good option in patients in whom an intra-aortic stent precludes local repair.

Although postoperative upper body blood pressure dropped to normal or near normal values in all patients, anti-hypertensive medication was usually continued for 6–8 weeks postoperatively to prevent paradoxical hypertension, occurring in up to 19% [6].

This small series demonstrates that extra-anatomic aortic bypass achieves excellent results in selected patients needing surgery because of primary or recurrent narrowing of the aortic arch or descending aorta. In our opinion, it is the procedure of choice for redo operations of complex forms of descending aortic disease. This technique may also be considered in patients with severe diffuse hypoplasia of the aortic arch and a long coarctation, to avoid the use of deep hypothermic circulatory arrest necessary for local repair. Close follow-up of these patients is necessary because of possible long-term complications.

References

Appendix A. ICVTS on-line discussion

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