Late complications of coarctation of the aorta

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Abstract

Background: The mechanism of the late complications after coarctation repair remains unclear, and this common congenital heart disease affects patients and perplexes physicians in terms of prevention and treatment.

Methods: From 2004 to 2008, 13 patients (1 adolescent and 12 adults) with repaired or unrepaired coarctation of the aorta were operated on in this department due to valve disorder or aortic aneurysm.

Results: The late complications were mitral and tricuspid regurgitation with congestive heart failure in 1, aortic valve disorder in 4, ascending aortic aneurysm in 3, saccular arch aneurysm in 1, and pseudoaneurysm in ascending, at isthmus, and descending aorta (Ortner’s syndrome) in 1 patient each, respectively. Recoarctation occurred in 3 (25%) patients, 23, 29, and 36 years after coarctation repair. One patient had persistent hypertension.

Conclusions: Patients with repaired coarctation of the aorta may eventually develop late complications including valve disorders, aortic aneurysm or pseudoaneurysm, in adolescence or adulthood, especially in the patient population associated with bicuspid aortic valve or complex congenital heart defects. Patients with unrepaired coarctation of the aorta, who can live to adulthood easily, may have a lack of associated anomalies. A regular follow-up is recommended for the patients with coarctation of the aorta in order to have a full-scope observation and prompt treatment when necessary. (Cardiol J 2008; 15: 517–524)

Key words: coarctation of the aorta, complications, mechanism

Introduction

Clinical observations have disclosed a typically higher incidence of late complications after coarctation repair when bicuspid aortic valve was present [1]. Little is known of the etiology of these complications in patients with coarctation of the aorta [2]. Late cardiovascular complications, such as valve disorders, aortic aneurysm or pseudoaneurysm, and hypertension always represent a danger to these patients in adolescence or adulthood, and challenge physicians in terms of effective prevention and treatment. We present 13 consecutive patients and discuss the mechanism of their late complications.

Methods

From January 2004 to July 2008, 13 patients (1 adolescent and 12 adults) with repaired or unrepaired coarctation of the aorta were operated on in this department due to valve disorder or aortic aneurysm. There were 8 males and 5 females. Twelve of them had coarctation repaired at an early age, and 1 did not. Their ages were 8.06 ± 9.57 years (range, 7 days – 30 years) at coarctation repair, and 33.25 ± 10.70 (17–48) years at current operation, with an interval between the two major operations of 25.15 ± 9.05 (1.75–47) years. Their clinical features are listed in Table 1.
<table>
<thead>
<tr>
<th>Case</th>
<th>Sex</th>
<th>Age (years)</th>
<th>Age at coarctation repair</th>
<th>Interval between coarctation and current operations (years)</th>
<th>Associated congenital disorders</th>
<th>Symptoms</th>
<th>Acquired disorders</th>
<th>History of operations other than coarctation repair</th>
<th>Current surgical indications</th>
<th>Surgery</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>25</td>
<td>7 days</td>
<td>25</td>
<td>Pulmonary stenosis, ventricular septal defect, atrial septal defect, nonobstructive cor triatriatum</td>
<td>Congestive heart failure</td>
<td>MR</td>
<td>Pulmonary artery banding and debanding, ventricular septal defect closure, atrial septal defect closure</td>
<td>MR, tricuspid regurgitation</td>
<td>Mitral valve replacement, tricuspid valve replacement, De Vega annuloplasty</td>
<td>Progressive heart failure in spite of inotropic support, ECMO, died 11 days after operation</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>31</td>
<td>29 years</td>
<td>1.75</td>
<td>Bicuspid aortic valve</td>
<td>Hypertension, infective endocarditis, AR, mild MR, right hand paresis due to septic emboli</td>
<td>AR</td>
<td></td>
<td></td>
<td>Aortic valve replacement</td>
<td>Survived</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>17</td>
<td>3 days</td>
<td>17</td>
<td>Bicuspid aortic valve, AR</td>
<td>Syncope</td>
<td>Balloon aortoventricular loplasty</td>
<td>AR + AS</td>
<td></td>
<td>Ross procedure</td>
<td>Survived</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>28</td>
<td>6 years</td>
<td>22</td>
<td>Bicuspid aortic valve</td>
<td>Ascending aortic aneurysm</td>
<td>Ascending aortic aneurysm</td>
<td></td>
<td>Aortic valve replacement + ascending aorta replacement + sino-tubular junction tailoring</td>
<td>Survived</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>23</td>
<td>8 days</td>
<td>23</td>
<td>Bicuspid aortic valve</td>
<td>Mild recoarctation, severe AS, moderate AR</td>
<td>Repair of recoarctation of aorta, aortic balloon valvuloplasty</td>
<td>AR + AS</td>
<td></td>
<td>Aortic valve replacement + manugian enlargement of the mitral valve anulus + aortic root with bovine pericardial patch</td>
<td>Survived</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>34</td>
<td>8 months</td>
<td>33</td>
<td>Bicuspid aortic valve</td>
<td>AS</td>
<td>Aortic valvuloplasty</td>
<td>AS</td>
<td></td>
<td>Aortic valve replacement</td>
<td>Survived</td>
</tr>
<tr>
<td>Case</td>
<td>Sex</td>
<td>Age</td>
<td>Age at coarctation repair</td>
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<td>Associated congenital disorders</td>
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<td>Current surgical indications</td>
<td>Surgery</td>
<td>Outcome</td>
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<tr>
<td>7</td>
<td>M</td>
<td>49</td>
<td>20</td>
<td>29</td>
<td>Bicuspid aortic valve</td>
<td>Fever and cough</td>
<td>AS, ascending aortic aneurysm, recoarctation</td>
<td>AS, ascending aortic aneurysm</td>
<td>Aortic valve replacement + ascending aorta replacement</td>
<td>Survived</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>41</td>
<td>9</td>
<td>32</td>
<td>Turner’s syndrome</td>
<td></td>
<td>Hypothyroidism, paroxysmal atrial fibrillation, paravalvular leak of prosthetic valve</td>
<td>Aortic valve replacement</td>
<td>Replacement of the aortic root and ascending aorta with 30 mm Valsalva Dacron graft and tricuspid annuloplasty</td>
<td>Postoperative, wide complex tachycardia, survived</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>M</td>
<td>28</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Aortic arch aneurysm — saccular type</td>
<td>Aortic arch replacement</td>
<td>Aortic arch replacement</td>
<td>Survived</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>M</td>
<td>33</td>
<td>3</td>
<td>30</td>
<td>Bicuspid aortic valve</td>
<td>Hoarseness</td>
<td>Descending aortic pseudo-aneurysm</td>
<td>Descending aortic pseudo-aneurysm (Ortner’s syndrome)</td>
<td>Descending aortic pseudo-aneurysmectomy</td>
<td>Survived</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>M</td>
<td>32</td>
<td>7</td>
<td>25</td>
<td>Bicuspid aortic valve</td>
<td>Shortness of breath</td>
<td>Severe AR, mediastinitis, complete atrioventricular block</td>
<td>Ascending aorta replacement, repair and reimplantation of native aortic valve, replacement of infected aortic graft with homograft, permanent pacemaker implantation</td>
<td>Bentall operation</td>
<td>Survived</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>F</td>
<td>55</td>
<td>19</td>
<td>36</td>
<td>Bicuspid aortic valve</td>
<td>Fatigue, short of breath</td>
<td>Pseudo-aneurysm at aortic isthmus</td>
<td>Stenting for recoarctation</td>
<td>Aortic valve replacement + ascending aorta replacement</td>
<td>Survived</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>M</td>
<td>31</td>
<td>3</td>
<td>28</td>
<td>Bicuspid aortic valve</td>
<td>Asymptomatic mild AR</td>
<td>Ascending aortic aneurysm, mild AR</td>
<td>Ascending aortic aneurysm</td>
<td>Ascending aorta replacement + reimplantation of the aortic valve</td>
<td>Survived</td>
<td></td>
</tr>
</tbody>
</table>

The study was approved by the local bioethical committee and all patients gave their informed consent.

**Results**

Bicuspid aortic valve was present in 10 (76.9%) of the 13 patients (1 of them associated with congenital aortic stenosis), Turner’s syndrome in 1 (7.7%) patient (whose native aortic valve was unknown), and multiple congenital heart defects (pulmonary stenosis, ventricular and atrial septal defects, and cor triatrium) in 1 patient (7.7%). Seven of the nine patients (77.8%) with a bicuspid aortic valve had aortic valve disorder (aortic stenosis in 2, aortic regurgitation in 3, and mixed stenosis and regurgitation in 2 patients), and one of them had a history of infective endocarditis with subsequent severe aortic regurgitation. Patients with isolated or mixed aortic stenosis had a decreased pressure gradient and flow velocity across the aortic valve, postoperatively (Fig. 1). In the patient whose coarctation was unrepaired, no additional congenital defect was present.

The late complications were mitral and tricuspid regurgitation with congestive heart failure was seen in 1 patient, aortic valve disorder in 4 patients, ascending aortic aneurysm in 4 patients, saccular arch aneurysm in 1 patient, and pseudoaneurysms in ascending, at isthmus, and descending aorta (Ortner’s syndrome) in 1 patient each, respectively. Recoarctation occurred in 3 (25%) patients, 23, 29, and 36 years after coarctation repair. Severe pulmonary hypertension was present in 1 patient, and mild pulmonary hypertension was present in another. One patient had persistent hypertension. All patients showed normal blood pressure (Fig. 2), with 7 of them on regular antihypertensive treatment.

Rib notching was noted in none of the posteroanterior roentgenograms of the 13 cases, including case 9 in whom coarctation was unrepaired. In case 1, the accessory atrium of cor triatrium was illustrated on axial computed tomography (Fig. 3). In cases 5, 7 and 12, recoarctation was visualized axially by computed tomography and 3-D reconstructed technique (Fig. 4). Pseudoaneurysms
of cases 10, 11 and 12 were finely demonstrated by conventional chest X-ray and axial computed tomo-
ography (Fig. 5). The saccular type arch aneurysm
of case 9 was visualized by 3-D reconstruction of
a Gadolinium-enhanced magnetic resonance an-
giography (Fig. 6).

Twelve (92.3%) patients survived the opera-
tion. Case 1 with cor triatrium associated pulmo-
nary hypertension had worsening congestive heart
failure after the operation. Even with inotropic sup-
port, her condition did not improve and had to be
aided by extracorporeal membrane oxygenation. She
deteriorated and died 11 days after the operation.

**Discussion**

In spite of effective repair of coarctation of the
aorta, the exercise capacity of adults after surgical
repair is reduced, and hypertension and early coro-
nary artery disease might occur [3]. Other compli-
cations such as aortic dilation developed in 7–26.1%
of the patients after coarctation repair [4, 5]. Mechani-
sms of the occurrence of aortic aneurysm, dissection,
or rupture after coarctation repair are unknown [2].
Clinical evaluations revealed that the presence of bi-
cuspid aortic valve, aortic dilation and hypertension
might contribute to its development [2, 6, 7].
Bicuspid aortic valve was present in 63–85% of the patients with coarctation of the aorta [5, 8]. We noted that 76.9% of our patients had an associated bicuspid aortic valve. In addition, we suspect that the patient who had undergone an aortic valve replacement also had a bicuspid aortic valve. Ciotti et al. [1] found patients with bicuspid aortic valve and coarctation of the aorta were less affected by valve disorder; however 77.8% of bicuspid aortic valve patients had valve disorder in this patient setting.

Other associated disorders may also contribute to the development of the complications late after coarctation repair. Ciotti et al. [1] reported that syndromes including Turner’s syndrome were associated with bicuspid aortic valve in 4.2% of patients. Mazzanti and Cacciari [9] noted that the prevalence of coarctation of the aorta was 6.9% in 594 Turner’s syndrome patients.

The incidences of late complications of coarctation varied, which were usually related to the associated cardiovascular disorders [2]. Late aortic complications were infrequent, with aortic aneurysm or dissection in 1%, and residual or recurrent coarctation in 6–10% of the operated patients [2]. Follow-up studies revealed the recoarctation rates were 9.5%, 15%, 16.3% and 41% in a group of 30 patients, although the operation rate for recoarctation was low (5.8%) [4, 5, 10, 11].

**Figure 6.** The saccular type arch aneurysm (upper arrow) and the unrepaired coarctation (lower arrow) were well visualized on sagittal view computed tomography (A), and 3-D reconstruction of a Gadolinium-enhanced magnetic resonance angiography (B, C).
Mechanisms that are responsible for the development of aortic pseudoaneurysms include infection, poor anastomotic technique, and intrinsic aortic disease [12]. Multivariate analysis demonstrated that bicuspid aortic valve, use of patch graft and advanced age at coarctation repair were independent predictors of pseudoaneurysm formation [6]. Santini et al. [12] noted cystic medial changes of the elastic fibers in the aortic specimen of a patient with ascending aortic pseudoaneurysm after bicuspid aortic valve replacement. Recently, Carr et al. [13] disclosed that pseudoaneurysm formation after coarctation repair was due to a commonly used silk suture in the previous era, whereas currently monofilament and polypropylene could lead to a relatively low incidence of pseudoaneurysm.

Mitrail disorder was rarely associated with coarctation of the aorta. Toro-Salazar et al. [5] and Ciotti et al. [1] noted incidences of 2.2% and 12.9%, respectively, of mitral disorders in patients with coarctation of the aorta, which were typically congenital abnormalities. In comparison, the mitral disorder of case 1 of the present patient group apparently resulted from complex congenital heart defects and severe pulmonary hypertension rather than induced directly by coarctation of the aorta.

Some authors [4, 5, 10] reported that 34.8–46% of patients with coarctation developed hypertension after surgical repair, although some of these patients were receiving antihypertensives. Continued hypertension may be due to residual coarctation in some patients, but without anatomic etiology in others, as described by Clark et al. [2]. Aortic arch has 3 geometries: gothic, crenel and normal. Gothic geometry of the aortic arch is associated with resting hypertension in patients with coarctation repair [14].

To gain a detailed record of postoperative alterations after aortic coarctation checkups at regular intervals are necessary. High-sensitive procedures, e.g. echocardiography, computed tomography, magnetic resonance imaging and intracardiac catheter are suitable means [15]. In diagnosing coarctation and recoarctation, oblique sagittal cine images could display the coarctation site. Gadolinium-enhanced magnetic resonance angiography was the best technique to show the entire thoracic aorta [16]. Collateral vessels could be better delineated with magnetic resonance imaging, while intracardiac anatomy and ventricular function could be better visualized with echocardiography [17].

Endovascular therapy of coarctation may lead to a restenosis of 6–40%. In comparison, surgery could be complicated by 9–18% restenosis [13]. In case of recoarctation the endovascular approach has to be recommended due to its positive short-term results. Long-term results remain to be seen [18, 19].

The morbidity of coarctation of the aorta remained higher, with only 31% free of cardiovascular morbidity at long-term follow-up [10]. The patients’ survival rates were 95%, 89%, 82% and 79% at 10-, 20-, 30- and 40-year follow-up, respectively [5].

Conclusions

Patients with repaired coarctation of the aorta may eventually develop late complications including valve disorders, aortic aneurysm or pseudoaneurysm, in adolescence or adulthood; this is especially true in the patient population associated with bicuspid aortic valve or complex congenital heart defects. Patients with unrepaired coarctation of the aorta who can live to adulthood easily may have a lack of associated anomalies. A regular follow-up is recommended for the patients with coarctation of the aorta in order to have a full-scope observation and prompt treatment when necessary.

Acknowledgements

The authors do not report any conflict of interest regarding this work.

References