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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

Editorial p. 491

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.

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Table 1. Clinical features.

Case	Sex	Age	Age at coarctation repair	Interval between coarctation and current operations (years)	Associated congenital disorders	Symptoms	Acquired disorders	History of operations other than coarctation repair	Current surgical indications	Surgery	Outcome
1	F	25	7 days	25	Pulmonary stenosis, ventricular septal defect, atrial septal defect, nonobstructive cor triatriatum	Congestive heart failure	MR	Pulmonary artery banding and debanding, ventricular septal defect closure, atrial septal defect closure	MR, tricuspid regurgitation	Mitral valve replacement, tricuspid De Vega annuloplasty	Progressive heart failure in spite of inotropic support, ECMO, died 11 days after operation
2	F	31	29 years	1.75	Bicuspid aortic valve		Hypertension, infective endocarditis, AR, mild MR, right hand paresis due to septic emboli		AR	Aortic valve replacement	Survived
3	M	17	3 days	17	Bicuspid aortic valve, AR	Syncope	Left ventricular hypertrophy, ventricular fibrillation resuscitated successfully	Balloon aortovalvuloplasty	AR + AS	Ross procedure	Survived
4	M	28	6 years	22	Bicuspid aortic valve		Ascending aortic aneurysm		Ascending aortic aneurysm	Aortic valve replacement + ascending aorta replacement + sinotubular junction tailoring	Survived
5	F	23	8 days	23	Bicuspid aortic valve		Mild recoarctation, severe AS, moderate AR	Repair of recoarctation of aorta, aortic baloon valvuloplasty	AR + AS	Aortic valve replacement + manugian enlargement of the mitral valve annulus + aortic root with bovine pericardial patch	Survived
6	M	34	8 months	33	Bicuspid aortic valve		AS	Aortic valvuloplasty	AS	Aortic valve replacement	Survived

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Table 1. cont. Clinical features.

Case	Sex	Age	Age at coarctation repair	Interval between coarctation and current operations (years)	Associated congenital disorders	Symptoms	Acquired disorders	History of operations other than coarctation repair	Current surgical indications	Surgery	Outcome
7	M	49	20 years	29	Bicuspid aortic valve	Fever and cough	AS, ascending aortic aneurysm, recoarctation		AS, ascending aortic aneurysm	Aortic valve replacement + ascending aorta replacement	Survived
8	F	41	9 years	32	Turner's syndrome		Hypothyroidism, paroxysmal atrial fibrillation, paravalvular leak of prosthetic valve	Aortic valve replacement	Ascending aorta aneurysm, paravalvular leak of prosthetic valve	Replacement of the aortic root and ascending aorta with 30 mm Valsalva Dacron graft and tricuspid annuloplasty	Postoperative, wide complex tachycardia, survived
9	M	28					Aortic arch aneurysm — saccular type		Aortic arch aneurysm — saccular type	Aortic arch replacement	Survived
10	M	33	3 years	30	Bicuspid aortic valve	Hoarseness	Descending aortic pseudoaneurysm	Aortic valvuloplasty	Descending aortic pseudoaneurysm (Ortner's syndrome)	Descending aortic pseudoaneurysmectomy	Survived
11	M	32	7 years	25	Bicuspid aortic valve	Shortness of breath	Severe AR, mediastinitis, complete atrioventricular block	Ascending aorta replacement, repair and reimplantation of native aortic valve, replacement of infected aortic graft with homograft, permanent pacemaker implantation	Ascending aortic pseudoaneurysm	Bentall operation	Survived
12	F	55	19 years	36	Bicuspid aortic valve	Fatigue, short of breath	Pseudoaneurysm at aortic isthmus	Stenting for recoarctation	Pseudoaneurysm at aortic isthmus	Aortic valve replacement + ascending aorta replacement	Survived
13	M	31	3 years	28	Bicuspid aortic valve		Ascending aortic, aneurysm, mild AR		Ascending aortic aneurysm, mild AR	Ascending aorta replacement + resuspension of the aortic valve	Survived

F – female, M — male, ECMO — extracarporeal membrane oxygenation, AS — aortic stenosis, AR — aortic regurgitation, MR — mitral regurgitation

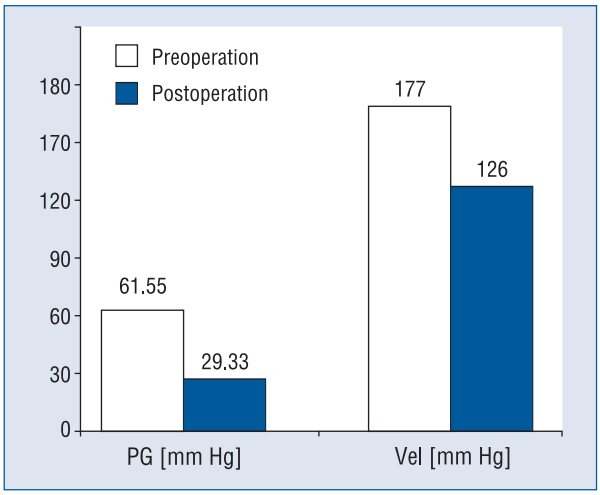


Figure 1. Pressure gradient (PG) and flow velocity (Vel) across the aortic valve were significantly decreased postoperatively in patients with isolated or mixed aortic stenosis.

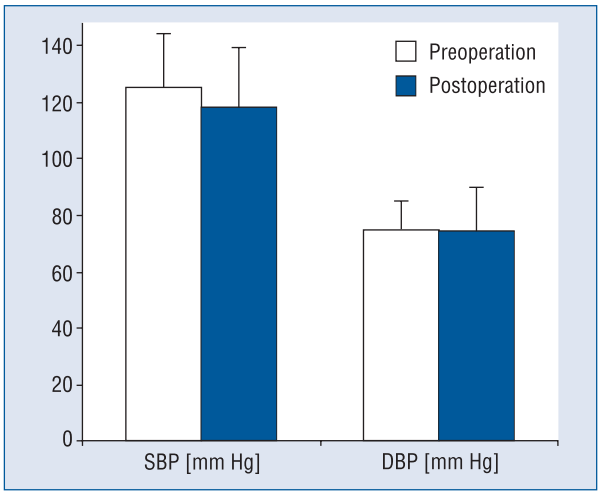


Figure 2. Systolic (SBP) and diastolic (DBP) blood pressure of patients with coarctation of the aorta at admission and at discharge.

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

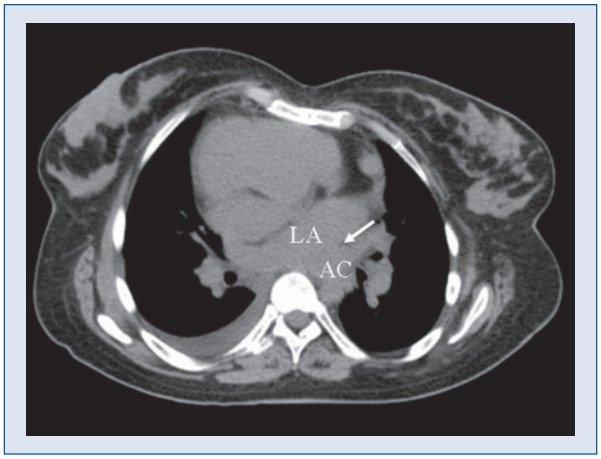


Figure 3. The accessory atrium of cor triatrium was illustrated on axial computed tomography. The wide opening (arrow) could be seen between the left atrium (LA) and the accessory chamber (AC).

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

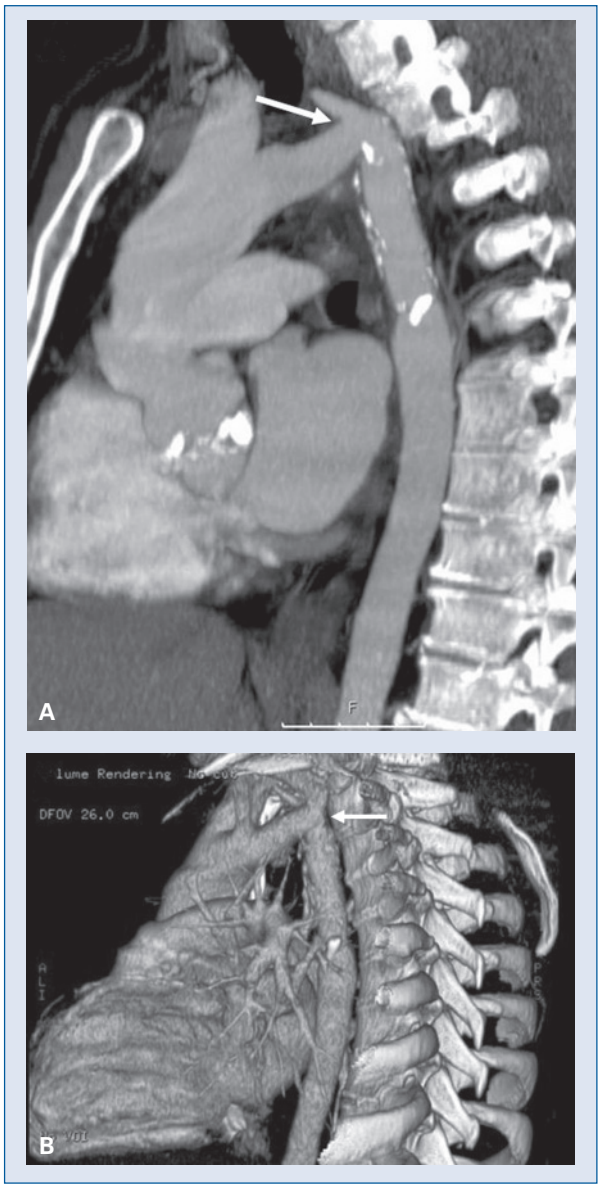


Figure 4. Sagittal computed tomography (**A**) and 3-D reconstructed technique (**B**) illustrated the recoarctation (arrow).

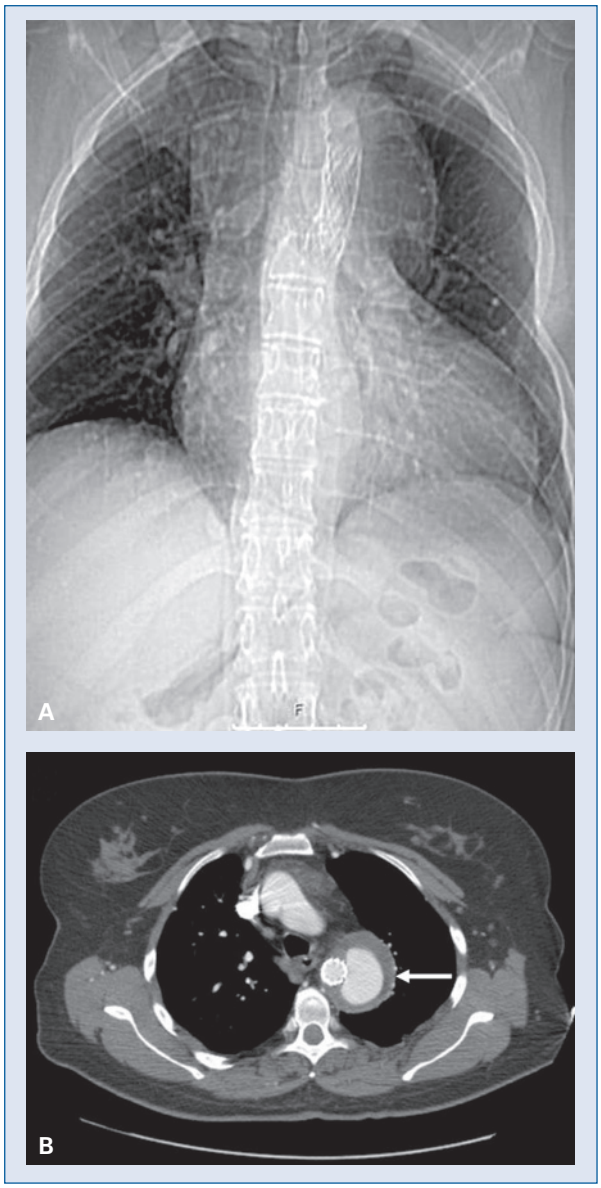


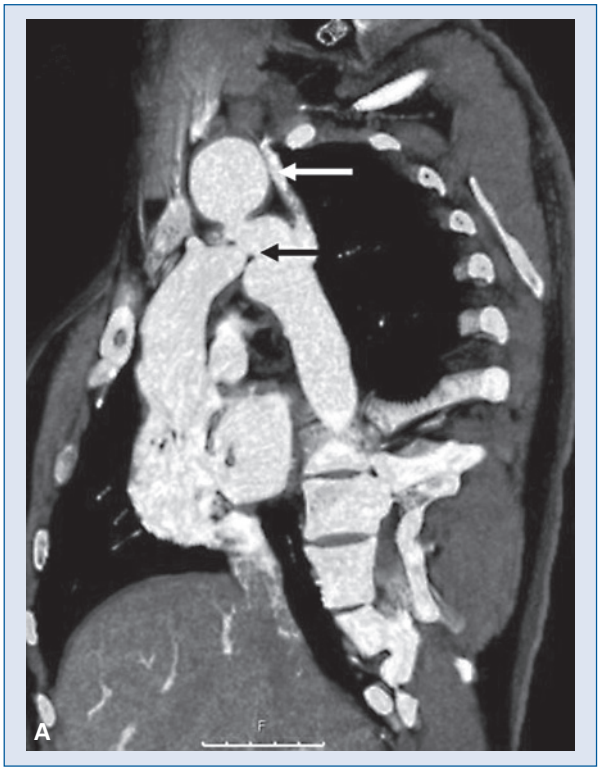
Figure 5. Chest X-ray (**A**) and axial computed tomography (**B**) showing aortic pseudoaneurysm at isthmus.

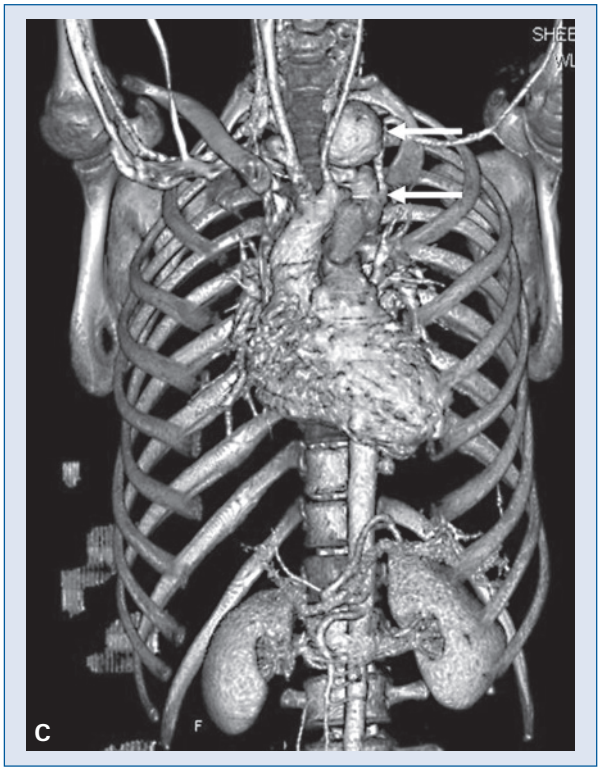
of cases 10, 11 and 12 were finely demonstrated by conventional chest X-ray and axial computed tomography (Fig. 5). The saccular type arch aneurysm of case 9 was visualized by 3-D reconstruction of a Gadolinium-enhanced magnetic resonance angiography (Fig. 6).

Twelve (92.3%) patients survived the operation. Case 1 with cor triatrium associated pulmonary hypertension had worsening congestive heart failure after the operation. Even with inotropic support, her condition did not improve and had to be aided by extracarporeal 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 coronary artery disease might occur [3]. Other complications such as aortic dilation developed in 7–26.1% of the patients after coarctation repair [4, 5]. Mechanisms of the occurrence of aortic aneurysm, dissection, or rupture after coarctation repair are unknown [2]. Clinical evaluations revealed that the presence of bicuspid aortic valve, aortic dilation and hypertension might contribute to its development [2, 6, 7].





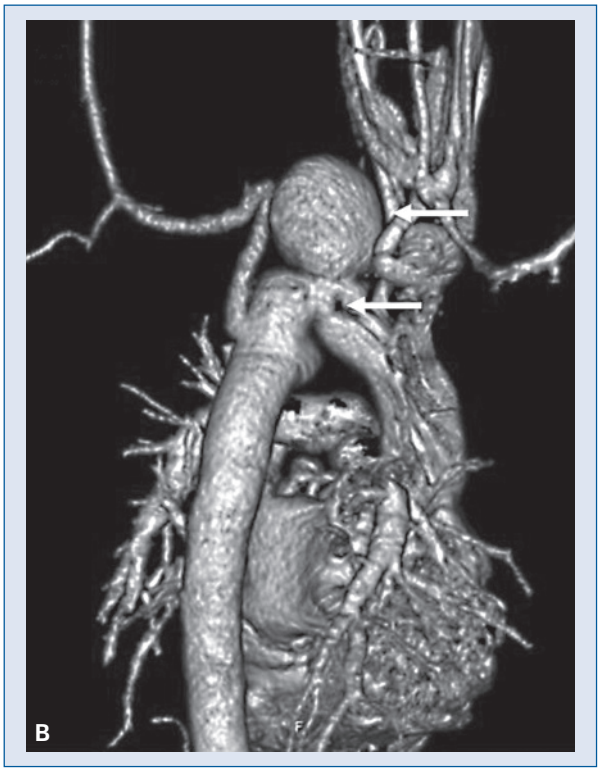


Figure 6. The saccular type arch aneurysm (upper arrow) and the unrepaired coarctation (lower arrow) were well visualized on sagittal view computed tomography (**A**), and 3-D reconstruction of a Gadolinium-enhanced magnetic resonance angiography (**B**, **C**).

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].

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.

Mitral 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

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