Selection of a Surgical Treatment Approach for Aortic Coarctation in Adolescents and Adults

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Purpose: Coarctation of the aorta (CoA) in adolescents and adults is relatively rare. Several operative techniques for CoA in adolescents and adults have been reported, but there is still no consensus. This study aims to highlight the use of individual patient characteristics to select optimal treatment strategies for CoA in adolescents and adults.

Methods: Surgical repair of CoA was performed in five patients (mean age: 34 ± 14 years, range: 13–58 years). All patients had primary CoA, and one had aneurysm above the CoA. One patient had undergone previous aortic valve replacement (AVR) and graft replacement of the ascending aorta. One patient underwent resection of the coarctation without cardiopulmonary bypass (CPB) followed by direct end-to-end anastomosis. Three patients underwent CoA resection with an interposition graft through a lateral thoracotomy with partial CPB. One patient underwent AVR with extra-anatomical bypass (ascending–descending aorta).

Results: No in-hospital deaths occurred, and there were no complications. During the follow-up period, there has been no recurrence of CoA.

Conclusion: CoA in adolescents and adults is associated with different issues from those encountered in infant patients, and comprehensive surgery should be performed in all cases.

Keywords: coarctation of the aorta, adolescents and adults, surgical treatment

Introduction

Coarctation of the aorta (CoA) is a common congenital heart disease defined as congenital stenosis of the aorta. CoA accounts for 57% of all congenital heart disease cases, with an incidence of approximately 3 cases per 10,000 births. CoA is almost always diagnosed in neonatal and infant patients, and congenital heart surgery for CoA is well established in infant patients. Treatment of adult CoA is challenging due to complication with other cardiac diseases, vascular abnormalities such as aneurysm, and possible CoA recurrence. Therefore, several operative techniques exist for CoA repair. Here, we report our experience with five cases of CoA in adolescents and adults.

Materials and Methods

Study population

Between 2006 and 2016, five adolescent and adult patients underwent CoA repair. Two patients were males, and three were females (mean age: 34 ± 14 years, range: 13–58 years). Preoperative findings are summarized in Table 1. Only one case had dyspnea. Four cases were complicated by bicuspid aortic valves and hypertension, one case was complicated by Turner syndrome.
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Two cases were complicated by other cardiac disease. All patients underwent primary repair for CoA, and the CoA was located in the region of the aortic isthmus in all patients. One patient had aneurysmal dilatation of the aorta above the aortic narrowing at the isthmus without a substantial pressure gradient (pseudo-coarctation).

**Operative procedure**

In four patients (No. 1–4), standard posterolateral thoracotomy was performed after insertion of a double-lumen endotracheal tube, while median sternotomy was performed in one patient (No. 5). Four patients (No. 2–5) were treated for CoA using cardiopulmonary bypass (CPB). In three patients (No. 2–4), partial CPB from the femoral vein to the femoral artery was used. Patient No. 5 was treated with full CPB. In all patients (CPB and non-CPB), surgery was performed at normal room temperature.

In patient No. 1, the length of abnormal aortic wall was approximately 2 cm, and operative femoral blood pressure decreased mildly (pre-clamp: 70 mmHg, post-clamp: 65 mmHg) in a tentative aortic clamping trial. Then, patient No. 1 underwent resection of the CoA without using CPB, and a direct end-to-end anastomosis was performed (Fig. 1). Patient No. 2 underwent CoA resection with interposition graft. Patient No. 3 suffered from CoA accompanied by an arterial aneurysm in the aortic arch and underwent partial graft replacement of the aortic arch with CoA resection through posterolateral thoracotomy (Fig. 2). Patients No. 4 and 5 suffered from concomitant cardiac disease and underwent two-stage and single-stage surgeries, respectively. Patient No. 4 first underwent an aortic valve replacement (AVR) and graft replacement of the ascending thoracic aorta. CoA resection with interposition graft was subsequently performed via posterolateral thoracotomy. In patient No. 5, AVR with extra-anatomical bypass (ascending–descending aorta) was performed (Fig. 3). The graft was passed through an incision on the posterior part of the pericardium and positioned along the right ventricle.

**Results**

No in-hospital deaths occurred, and there were no complications such as reoperation for bleeding, renal failure, spinal cord injury, stroke, myocardial failure, left recurrent laryngeal nerve injury, or chylothorax. For the three patients treated for CoA using partial CPB, mean CPB time was 70 ± 18 min. For the patient treated with total CPB, CPB time was 251 min. The mean operative time was 436 ± 138.9 min. The mean stay in the intensive care unit was 3.8 ± 1.9 days (range: 2–7 days), and mean total hospital stay was 20.8 ± 8.9 days.

<table>
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<th>Table 1 Preoperative findings</th>
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<tr>
<td>Patient No</td>
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<td>Age (years)</td>
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<tr>
<td>Sex</td>
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<tr>
<td>BSA (m²)</td>
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<tr>
<td>Hypertension</td>
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<td>Bicuspid valve</td>
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<td>Aneurysm</td>
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<td>PG across CoA (mmHg)</td>
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<td>Other complication</td>
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CoA: Coarctation of the aorta; Asc.Ao.Rep: ascending aorta replacement; BSA: body surface area; PG: pressure gradient; AR: aortic regurgitation.

Fig. 1 Three-dimensional CT. (A) Preoperative view. (B) Postoperative view. A direct end-to-end anastomosis was performed. CT: computed tomography.
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(range: 6–27 days). Operative and postoperative findings are summarized in Table 2.

During the follow-up period (75 ± 49 months), there has been no recurrence of CoA.

Discussion

Congenital heart surgery for CoA is well established in infant patients. However, when patients present CoA later in life, the vessels are much more friable, and the intercostal arteries are larger and more easily damaged, making dissection potentially more hazardous. In addition, some cases may be complicated by additional cardiovascular disease, previous surgery, and/or CoA
recurrence (10%–44.7%). Therefore, several operative techniques have been proposed for CoA repair in adolescents and adults depending on individual patient circumstances. The surgical procedures can be roughly divided into three categories: resection with end-to-end anastomosis, CoA resection with interposition graft, and extra-anatomical bypass without CoA resection.

CoA resection with end-to-end anastomosis was performed by Crafoord and Gross in 1945 through a lateral thoracotomy. CoA resection with end-to-end anastomosis has the advantage of resecting the abnormal aortic wall with ductal tissue rather than using a prosthetic graft and is therefore considered the ideal procedure. In neonate patients, CoA resection with end-to-end anastomosis is easily performed after extended resection, but in adolescents and adults, CoA resection with end-to-end anastomosis is more technically challenging due to the thickness of the aortic wall, difficulties in aortic arch mobilization, severe calcification, and large collateral arteries.

We performed CoA resection with end-to-end anastomosis in patient No. 1 because the aortic arch distal to the left carotid artery and the descending aorta were easily mobilized from the root of the neck to a distance well below the CoA. Furthermore, since patient No. 1 is expected to continue growing, we wanted to avoid using a prosthetic graft. Although this procedure is occasionally performed in adults, we believe that its indication is limited (e.g., young cases, short-segment CoA) because its difficulty increases in patients who present later in life. In addition, Sandro et al. reported that an intraoperative femoral blood pressure of <50 mmHg on tentative aortic clamping trial necessitates repair of the CoA using CPB to protect the spine and lower body. We performed the operation without CPB in patient No. 1 because intraoperative femoral blood pressure was 65 mmHg.

CoA resection with interposition graft as described by Gross was performed via lateral thoracotomy. This procedure is recommended particularly in cases of long-segment CoA and CoA with aneurysm formation. In adults, this procedure may be preferable for many surgeons because the disadvantage of the tube graft not growing with the patient is not a concern. Moreover, this procedure can be used to perform resection of CoA and abnormal vessel features such as aneurysm. On the other hand, its major disadvantages are the longer cross-clamp time for two anastomoses. CPB was used in patient No. 2 to avoid injury to the spinal cord and visceral organs caused by distal ischemia related to the longer cross-clamp time.

Our three cases (patient No. 2, No. 3, and No. 5) underwent CoA resection with interposition graft using partial CPB. Other studies have reported avoiding distal ischemia using a Gott shunt or intraluminal shunt. In more complicated cases, the use of total CPB and hypothermic circulatory arrest for minimal dissection has been recommended as a surgical procedure for CoA, after which CoA resection with interposition graft or CoA resection with end-to-end anastomosis is performed. Elkerdany et al. also reported good outcomes in adults after the treatment of isolated CoA by left subclavian-aortic bypass. This procedure avoids dissection through the hard adhesions between the CoA and surrounding organs; however, it cannot be used in the hypoplastic arch, and abnormal aortic wall remains.

In adults, we may also encounter the need to perform surgery for cases of CoA recurrence and CoA with other cardiac disease. In these cases, extra-anatomical bypass without CoA resection is recommended in some patients. Interposition grafting is difficult in recurrent CoA repair via left thoracotomy due to adhesion. Consequently, reoperation via left thoracotomy has a high incidence of postoperative complications, with reported mortality and morbidity rates of 0–8% and 50%, respectively. Since the approach for extra-anatomical bypass is performed via median sternotomy or right thoracotomy, it avoids the complications associated with repeated left thoracotomy. In addition, extra-anatomical bypass via median sternotomy can be performed even in cases where concomitant surgical repair of CoA and another cardiovascular disease is planned. Mesnildrey et al. reported that 37% of adult CoA cases presented with cardiopathy. In such cases, two-stage repair, single-stage repair with median sternotomy and left thoracotomy, and single-stage repair with extra-anatomical bypass have been reported. Among these procedures, single-stage repair with extra-anatomical bypass may lower both the associated risk and patient discomfort. In this procedure, proximal anastomosis is typically performed in the ascending thoracic aorta, but the appropriate site of distal anastomosis (abdominal aorta or descending thoracic aorta) is still under debate. Patient No. 5 was treated by AVR with extra-anatomical bypass (ascending–descending aorta) in a single-stage repair. We opted for single-stage repair in Patient No. 5 because Turner’s syndrome and cognitive dysfunction were observed in addition to heart disease. In Patient No. 5, computed tomography (CT) demonstrated aneurysm
formation distal to the CoA site preoperatively. However, the aneurysm diameter was only 40 mm, and to avoid high operative risk, we did not perform aneurysm resection with interposition graft in this patient. This patient is currently receiving follow-up care involving annual CT to monitor the aneurysm.

There were two possible routes of extracardiac bypass graft in the pericardium. In the route of the graft leading around the left aspect of the heart, the length of the graft was shorter, but there was a possibility of graft kinking and compression of surrounding structures. In the route of the graft leading around the right aspect of the heart, avoids the risk associated with re-sternotomy, as the graft is not near the sternum. Since patient No. 5 was young, we chose the route around the right aspect of the heart to allow for the possibility of re-sternotomy.

In patient No. 4, we first performed an AVR and graft replacement of the ascending thoracic aorta. CoA resection with interposition graft was subsequently performed via posterolateral thoracotomy. In this patient, it was necessary to treat aortic regurgitation (AR), aneurysm of the ascending thoracic aorta, and CoA. Considering the invasiveness to the patient, we performed two-stage repair intentionally. However, the first and second surgeries were separated by a 19-month period because the patient did not wish to undergo an additional procedure after becoming asymptomatic after the first operation. If possible, we recommend single-stage repair with extra-anatomical bypass.

In addition, endovascular therapy is becoming more widespread for the treatment of CoA. In 1982, transcatheter balloon angioplasty was initially reported by Singer et al. in infants. 23) Subsequently, this treatment has spread to adult patients, and good results have been reported using stents. However, due to the higher rates of recurrence and aneurysm formation associated with endovascular treatment, patients treated with this method require long-term follow-up. 24)

In summary, CoA resection using interposition graft with CPB is recommended for spinal cord and lower body protection in adult patients. However, in adolescents, resection with end-to-end anastomosis should also be considered if continued growth is expected. The use of CPB may not always be necessary because the end-to-end anastomosis ends in a short time, but CPB should be used when the intraoperative femoral blood pressure is <50 mmHg on tentative aortic clamping trial. In cases of CoA recurrence and CoA with other cardiac disease, extra-anatomical bypass without CoA resection should also be considered.

**Conclusion**

The treatment of CoA in adult patients presents different challenges to those encountered in infants. As the treatment of CoA has progressed, a range of treatments can result in excellent outcome if each patient’s individual circumstances (such as repeat operation, aneurysm formation, large collaterals, and cardiovascular pathology) are carefully considered preoperatively. As the number of cases included in this study is small, further research is necessary to establish the optimal treatment for CoA in adolescents and adults in the future.

**Disclosure Statement**

The authors declare no conflicts of interest.

**References**