One-Stage Correction of Complex Aortic Arch and Descending Aorta Dysplasia Combined Intracardiac Anomalies with a Median Incision

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Abstract

Objectives: A congenital aortic arch and descending aorta dysplasia with intracardiac anomalies are rare cardiovascular malformations, and there is some controversy regarding the ideal surgical response. Our objective is to report our experiences regarding the one-stage correction for these complex cardiovascular anomalies in adolescent and adults.

Methods: The clinical data were analyzed retrospectively from 49 patients who underwent surgery for these complex anomalies. Surgical treatments for aortic malformations include widening of the aortic arch and descending aorta in 31 patients (Widening group) and ascending aorta-Gore Tex graft-descending aorta bypass in 18 patients (Bypass group).

Results: There was one early death each in the Bypass group and the Widening group. There were 2 patients with early hoarseness and 2 patients with chylothorax in the Widening group. The change in the systolic pressure gradients between the upper and lower limbs was less than 10 mmHg in all patients within one year of follow-up. Conclusions Median sternotomy and concomitant surgical treatment for aortic arch and descending aorta dysplasia associated with intracardiac anomalies are safe and feasible. Gore-Tex vascular graft bypass was a good choice for patients with interrupted aortic arch, distal aortic arch dysplasia, or obvious vascular calcification in adolescents and adults.

Keywords: Aortic arch; Descending aorta dysplasia; Intracardiac abnormalities; Surgical treatments

Introduction

Congenital complex aortic arch with descending aorta dysplasia is a rare cardiovascular malformation. It is mainly divided into the following three types: simple aorta coarctation, aorta coarctation with isthmus hypoplasia, and aorta coarctation with aortic arch dysplasia. Simple aorta coarctation has good surgical outcomes. Interventional balloon dilation and stents can be used [1-7]. The other two types of aorta coarctation are complicated and have poor surgical outcomes. Treatment for patients with aorta coarctation associated with intracardiac anomalies is still controversial [7-9]. In this study, we describe and discuss our experiences with median sternotomy and concomitant surgical treatment against complex aortic arch and descending aortic dysplasia associated with intracardiac anomalies in forty-nine patients within a single institute.

Materials and Methods

Patients

Forty-nine patients were included in this study, consisting of 28 boys and men and 21 girls and women ranging in age from 12 to 33 years. The patients included 11 cases of interrupted aortic arch and 38 cases of aortic arch and descending aorta dysplasia (with coarctation segments longer than 4 cm). The intracardiac complications included 35 cases of ventricular septal defect, 5 cases of bicuspid aortic valve with stenosis, 4 cases of right ventricular outflow tract obstruction or double-chambered right ventricle, 2 cases of atrial septal defect, 1 case of pulmonary stenosis, 1 case of mitral stenosis, 1 case of aortic stenosis with mitral stenosis, and 30 cases of patent ductus arteriosus. Clinical manifestations included exertional dyspnea, shortness of breath, intermittent lower limb weakness, occasional chest pain, lower blood pressure in the lower limbs than in the upper limbs, thin and impalpable femoral artery and dorsalis pedis artery, systolic or continuous murmur at the left sternal border between the 3rd and 4th intercostal spaces, and loud P2.
seven patients showed differential cyanosis. Chest X-ray examination showed bilateral pulmonary congestion and prominent pulmonary artery segmentation. ECG showed left ventricular hypertrophy. Echocardiography showed corresponding heart malformations, which were confirmed by surgery (Table 1), and invisible descending aortas. CT aortic angiography clearly showed aortic arch and descending aortic dysplasia (Figure 1-4) and rich collateral circulation. Intraoperative exploration showed left ventricular dilation and hypertrophy, right ventricular enlargement, pulmonary artery thickening, diffusive coarctation at the distal aortic arch, descending aortic dilation after coarctation, and thin and fragile tissue and vessel walls. Each participant enrolled in our study was assigned to the Widening Group or Bypass Group based on their standard treatment directed by the surgeon. Our study was approved by the institutional research ethics committee of Xinqiao Hospital. All the participants in our study provided written informed consent. Additionally, we had obtained written informed consent from the statutory guardian of the minors enrolled in our study.

**Surgical approaches**

Surgery was performed under intravenous anesthesia. Surgical procedures included median sternotomy, longitudinal incision of the pericardium, and preparation of a pericardial patch. The patients were divided into two groups based on the surgical approaches widening group (widening of the aortic arch and descending aorta in 31 patients) and Bypass group (ascending aorta-Gore Tex graft-descending aorta bypass in 18 patients).

(1) In the Widening group comprising 31 patients with long aortic arches and descending aortic dysplasia, we carefully separated the coarctation segment of the descending aorta along the aortic arch from the normal sites, which were 2 cm to 3 cm above the coarctation segment. Once the separation was complete, the conventional cardiopulmonary bypass was set up. When extracorporeal circulation was cooled down to 18°C, and the subject underwent circulatory arrest, the coarctation segment was incised longitudinally, widened with blood vessel grafts and lined externally with strips of autologous pericardium to prevent blood vessel rupture. We also added one layer of strips of autologous pericardium to the lumens of the blood vessels to prevent post-operative bleeding. Next, we exhausted and restored circulation, warmed the patient up slowly, perfused the heart with cardiac arrest fluid, and completed the correction of the anomalies.

(2) In the Bypass group comprising 7 patients who had long distal...
warfarin for 3 months to prevent thrombosis. After surgery, all patients were given perfusion with 500 ml of arrest solution. This was completed in 3 min circulation and perfusion, we performed superior vena cava retrograde ductus arteriosus closure in 30 patients. Before the restoration of replacement in 1 patient, Bent all procedure in 1 patient, and patent in 2 patients, mitral valve replacement in 1 patient, multi-valve ventricular outflow tract in 5 patients, atrial septal defect correction in 35 patients, aortic arch dysplasia and rich collateral circulation in CTA and 11 patients who had interrupted aortic arches, we carefully separated the descending aorta from the lower end through the extra pleura, which was 3 cm to 4 cm above the normal area, set up the conventional cardiopulmonary bypass, placed the aortic cannula on the right side of the aorta and as close as possible to the aortic arch, reduced the patient’s systemic temperature to 18°C, and performed Gore-Tex bypass graft replacement with circulatory arrest. First, we performed end-to-side anastomosis between the Gore-Tex vascular grafts and normal descending aorta distal from the coarctation. The anastomosis was continuously sutured with a 5/0 prolene line and was lined and tightened with stripped autologous pericardium. We next restored perfusion with cold cardiac arrest solution, and corrected the intracardiac anomalies. Finally, we performed end-to-side anastomosis between another end of the Gore-Tex vascular graft and normal ascending aorta and restored aortic flow. In five patients in this group, the coarctation segment was too long for the normal part of the descending aorta to be fully exposed through extra pleura; thus, we opened the pleura, bypassed through the left lung using long Gortex vascular grafts, and performed end-to-side anastomosis with descending aorta above the diaphragm. The same procedure was used for the proximal end.

Concomitant surgical treatments for intracardiac anomalies included ventricular septal defect correction in 35 patients, aortic valve replacement in 4 patients, dregdging and widening of the right ventricular outflow tract in 5 patients, atrial septal defect correction in 2 patients, mitral valve replacement in 1 patient, multi-valve replacement in 1 patient, Bent all procedure in 1 patient, and patent ductus arteriosus closure in 30 patients. Before the restoration of circulation and perfusion, we performed superior vena cava retrograde perfusion with 500 ml of arrest solution. This was completed in 3 min to 5 mins to allow gas to be expelled and prevent air embolism in the brain and other vital organs. After surgery, all patients were given warfarin for 3 months to prevent thrombosis.

**Measurement of peripheral oxygen saturation**

Changes in the systolic pressure gradients between upper and lower limbs in 47 surviving patients before the operation, 1 day post-operation, 30 days post-operation, 6 months post-operation, and 1 year post-operation were analyzed.

**Statistical analysis**

The Statistical Package for the Social Sciences (SPSS) version 11.0 was utilized for to data analysis. The count data were assessed by χ² test. All data were reported as the means ± standard deviation. P<0.05 was considered statistically significant.

**Results**

The surgical approaches are listed in (Table 1). One patient died during the early stages of surgery due to low cardiac output syndrome in the Bypass group, and one patient died during the early stages of surgery due to respiratory function failure in the Widening group. No deaths occurred late in the two groups. There were 12 cases (38.71%) with complications in the Widening group, including 4 patients with low cardiac output syndrome, 2 patients with respiratory function failure, 2 with multi-organ failure, 2 with early hoarseness, and 2 with chylothorax. One patient required reoperation because of bleeding.

In the Bypass group, 5 patients (27.78%) with complications that were lower than those in the Widening group comprised 2 patients with low cardiac output syndrome, 1 patient with respiratory function failure and 1 patient required reoperation because of bleeding. All of them were cured and discharged after treatment. We performed follow-up for 3 months to 9 years, and all patients were satisfied with the outcome and had a good quality of life. The changes in the systolic pressure gradients between the upper and lower limbs within one year of surgery in 47 surviving patients (± s mmHg).

**Table 2:** Changes in the systolic pressure gradients between the upper and lower limbs within one year of surgery in 47 surviving patients (± s mmHg).

<table>
<thead>
<tr>
<th></th>
<th>Upper limb</th>
<th>Lower limb</th>
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<tbody>
<tr>
<td>Pre-operation</td>
<td>166.79 ± 15.93</td>
<td>167.89 ± 16.14</td>
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<tr>
<td>1 day post-operation</td>
<td>125.23 ± 10.78</td>
<td>129.11 ± 16.93</td>
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<tr>
<td>30 days post-operation</td>
<td>135.81 ± 17.83</td>
<td>138.22 ± 8.61</td>
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<tr>
<td>6 months post-operation</td>
<td>140.72 ± 10.10</td>
<td>137.67 ± 8.33</td>
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<tr>
<td>1 year post-operation</td>
<td>143.03 ± 11.72</td>
<td>139.89 ± 8.92</td>
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</table>

**Table 3:** Changes in the systolic blood pressure of the upper and lower limbs within one year of surgery in 47 surviving patients (± s mmHg).

<table>
<thead>
<tr>
<th></th>
<th>Upper limb</th>
<th>Lower limb</th>
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<tbody>
<tr>
<td></td>
<td>Widening Group (N=30)</td>
<td>Bypass Group (N=17)</td>
</tr>
<tr>
<td>Pre-operation</td>
<td>72.27 ± 14.94</td>
<td>72.61 ± 15.67</td>
</tr>
<tr>
<td>1 day post-operation</td>
<td>12.29 ± 5.90</td>
<td>12.89 ± 6.43</td>
</tr>
<tr>
<td>30 days post-operation</td>
<td>6.18 ± 6.24</td>
<td>6.11 ± 6.37</td>
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<tr>
<td>6 months post-operation</td>
<td>0.59 ± 7.00</td>
<td>-0.44 ± 6.75</td>
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<td>1 year post-operation</td>
<td>-1.68 ± 6.54</td>
<td>-2.72 ± 5.42</td>
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**Discussion**

Congenital complex aortic arch associated with descending aortic dysplasia is a rare cardiovascular malformation. It is divided into the following three types: simple aortic coarctation, aortic coarctation with isthmus hypoplasia, and aortic coarctation with aortic arch dysplasia. Surgical treatment for simple aortic coarctation is easy and
effective. Interventional balloon dilation and stents are commonly used. The other two types of aortic coarctation are complicated, and both surgical and interventional approaches are difficult to apply. It is more difficult to treat when they are associated with intracardiac anomalies, and treatment strategies are still controversial [7-11].

**Selection of surgical approach**

The conventional surgical approaches for simple aortic coarctation include left posterior lateral incision, excision of the coarctation segment through the fourth intercostal space, and end-to-end anastomosis. It can also be corrected with incision and widening at the coarctation segment, which has good exposure. This technique requires only that the surface of the body be cooled and does not require cardiopulmonary bypass or deep hypothermic circulatory arrest. It is simple, and the outcomes are good. Interventional balloon dilation and stents also have good effects [1-7].

Due to the complicated pathophysiology and haemodynamics, there is no standard surgical approach to aortic coarctation associated with complications such as isthmus hypoplasia, aorta arch dysplasia, and interrupted aorta arch or intracardiac anomalies [8]. Common surgical strategies include staging surgery, single-stage surgery with two incisions, and single-stage surgery with a median incision [11-13]. Staging surgery includes left posterior lateral incision to correct aortic anomalies and selective surgery for intracardiac anomalies. Single-stage surgery with two incisions includes left posterior lateral incision to correct aortic anomalies and concomitant correction of intracardiac anomalies after changing the posture of the patient. Both methods have good exposure in the surgical field. However, patients must undergo these painful and risky procedures twice. Single-stage surgery with median sternotomy has relatively poor exposure in the surgical field and poorer parameters than single-stage surgery with two incisions with respect to the cardiopulmonary bypass time, duration of ICU stay, mortality rate, and re-coarctation rate. However, it is the best choice for newborns and infants with aortic arch dysplasia [11-13]. It remains controversial whether this method is appropriate for adolescents and adults [12,13]. Our experiences suggest that, in adolescents and adults, the median sternotomy approach can be used to successfully complete the correction with fewer incisions and a low re-operation rate. It may also be easier for patients to accept.

**Surgical strategies**

Surgical treatment is the best approach for the treatment of descending aortic dysplasia associated with intracardiac anomalies. It improves the natural prognosis. However, no surgical strategy is universally accepted. In end-to-end anastomosis, there is usually insufficient vascular anastomosis, and anastomotic stricture may occur. Aneurysms may occur after patch widening [3,8,14].

In the present study, we chose surgical approaches in accordance with patient-specific conditions. For patients with aortic coarctation and large-area isthmus hypoplasia, patients with concomitant aortic arch dysplasia but mild coarctation, and young patients, we selected longitudinal incision and Gore-Tex vascular graft plus autologous pericardium to widen the narrow section of the aortic arch. Patch widening can reduce the risk of post-operative stenosis [8]. Lining with autologous pericardium can prevent aortic wall rupture and post-operative bleeding. Gore-Tex vascular grafts have good elasticity and plasticity and are less prone to post-operative aneurysm [15].

For patients with interrupted aortic arches, distal aortic arch dysplasia, long coarctation segments, or obvious vascular calcification and older children and adults, we adopted a Gore-Tex vascular graft bypass (Figure 1). First, we performed end-to-side anastomosis between the Gore-Tex vascular graft and normal descending aorta distal from the coarctation, and the anastomosis was continuously sutured with 5/0 prolene line, tightened and lined with strips of autologous pericardium. Next, we restored the cardiopulmonary bypass, allowed perfusion, and warmed up the blood. After expelling gas from the vascular graft and clamping the said graft, we performed end-to-side anastomosis between the Gore-Tex vascular graft and normal ascending aorta. We only performed end-to-side anastomosis between the vascular graft and normal descending aorta with circulatory arrest. This can decrease the total surgical time and facilitate good anastomosis. Lining with stripped autologous pericardium can prevent vessel rupture. Unlike widening between the descending aorta and distal aortic arch, complete separation of
the lateral and posterior walls of the aorta was unnecessary. This left a smaller wound area, a lower chance of chylothorax, and intra-operative bleeding due to the rich collateral circulation and vessel wall fragility. If the coarctation segment was too long for full exposure of the normal part of the descending aorta through the extra pleura, we opened the pleura, bypassed the left lung using long Gortex vascular grafts and performed end-to-side anastomosis above the diaphragm (Figure 2). In this way, we prevented deficits in the length and tension of the graft vessels and prevented poor outcomes caused by lung expansion. This procedure was used in all five patients in this group, to good effect. We then performed end-to-side anastomosis between the vascular graft and ascending aorta after restoring cardiopulmonary bypass and perfusion, warming the blood, and blocking the ascending aorta. This can reduce the circulatory arrest time and cardiopulmonary bypass time and reduce their effects on the body. End-to-side anastomosis allows the physician to choose large vessel grafts, which reduce the rate of stenosis and avoid the problem of matching. During cannulation of the ascending aorta, the cannulation should be placed to the right of the blood vessel and as close as possible to the aortic arch. This provides sufficient space to establish anastomosis between the vascular graft and ascending aorta. The correction of intracardiac anomalies can be performed after the lesion vessels are separated and the cardiopulmonary bypass is cooled down or after the aortic malformations are corrected and the cardiopulmonary bypass is warmed up. The process works well under either set of conditions. We performed this procedure in 18 patients, and it was successful in all cases.

In the present study, the changes in the systolic pressure gradients between the upper and lower limbs were less than 10 mmHg in all patients within one year of surgery, and there was no significant difference in the Gore-Tex vascular graft bypass group compared with that in the Patch widening group. It showed that the Gore-Tex vascular graft bypass was a good choice for patients with interrupted aortic arches, distal aortic arch dysplasia, long coarctation segments, or obvious vascular calcification for adolescents and adults.

Protection of nearby vital organs

In this study, chylothorax occurred in 3 patients, and early-onset hoarseness occurred in 2 patients, indicating the importance of protecting important neighboring tissues. The phrenic nerve, vagus nerve, and recurrent laryngeal nerve pass around the aortic arch and descending aorta; thus, close attention must be paid to prevent damage to these nerves. Electrocoagulation-blunt dissection should be used at low output intensity to prevent indirect thermal damage. If necessary, the nerves should be traced with a rubber strip. There are abundant amounts of lymphatic tissue around the aortic arch and descending aorta, and ligation should be used to reduce the chance of chylothorax [16-18].

Advantages of concomitant perfusion and cannulation of the ascending aorta and femoral artery

In adult patients, we performed concomitant perfusion and cannulation of the ascending aorta and femoral artery because of the weak tissue of the vessel walls and because of the long surgical time required for adults. We chose either upper body perfusion or lower body perfusion under circulatory arrest to shorten the duration of ischaemia of vital organs and reduce the damage to organ function. Because this process involves selective perfusion, the surgery can take as long as required for perfect correction of the deformity, reducing the chances of post-operative bleeding and other complications. In this case, we achieved perfect correction of anomalies and reduced post-operative bleeding. In addition, concomitant perfusion of the ascending aorta and femoral artery can evenly and quickly cool down the whole body. This prevents uneven cooling by a single perfusion and the adverse effects attributable to uneven cooling of the distal end of the coarctation. It works well in clinical settings.

Significance of retrograde perfusion of the superior vena cava

To completely expel gas, a superior vena cava retrograde perfusion was applied in patients. This technique can completely exhaust intravascular gas and prevent air embolism in the brain and other vital organs. The perfusion pressure should not be too high. Normally, it is less than 40 mmHg, and the perfusion volume is normally 500 ml. The perfusion should be performed within 3 min to 5 mins. After the retrograde perfusion, arterial perfusion was restored, and the body was warmed up slowly. No brain complications occurred in this group of patients.

Conclusion

In summary, a complex aorta arch and descending aorta dysplasia associated with intracardiac anomalies are complicated situations. Median sternotomy and concomitant surgical treatment are safe and feasible. A Gore-Tex vascular graft bypass was a good choice for patients with interrupted aortic arches, distal aortic arch dysplasia, long coarctation segments, or obvious vascular calcification for adolescents and adults. No recurrence was observed during follow-up, and no related complications occurred.

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References


