Surgical modification for preventing a gothic arch after aortic arch repair without the use of foreign material

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Abstract

OBJECTIVES: Systemic hypertension is the main late complication after arch reconstruction in patients with arch obstruction. Gothic arch geometry is suspected to be one of its possible causes. Accordingly, we evaluated here if a modified arch repair technique using an autologous pulmonary patch is effective in preventing gothic arch development.

METHODS: Fifty infants who underwent arch repair with either a modified (n = 17) or conventional (n = 33) technique between January 2006 and August 2012 by a single surgeon were retrospectively reviewed. Arch geometry was compared using three categories (gothic, crenel or roman), classified by the height/width (H/W) ratio and the arch angle measured in computed tomography.

RESULTS: No gothic arch geometry was observed in the modified group, whereas it was observed in 9 cases in the conventional group (P = 0.005). Moreover, reintervention for arch restenosis was performed only in the conventional group (n = 4; P = 0.29). No associated complications were observed, although the selective cerebral perfusion time was longer in the modified group than in the conventional group (28.5 ± 6.2 vs 17.1 ± 9.9 min; P < 0.001). Otherwise, there were no significant differences in clinical variables between the groups. The mean follow-up duration was 55.3 ± 26.7 months. Significant systemic hypertension was not observed in our study cohort.

CONCLUSIONS: Our modified technique was proven to be not only highly effective in preventing gothic arch geometry, but also as equally safe in terms of early clinical outcomes as conventional arch reconstruction techniques.

Keywords: Coarctation of aorta • Arch hypoplasia • Arch repair • Hypertension

INTRODUCTION

Neonatal one-stage total correction is now accepted as a standard treatment for patients with aortic arch obstruction with intracardiac anomalies [1–3]. Improved surgical techniques and perioperative care have decreased early operative mortality. However, systemic hypertension remains one of the major late complications of an aortic arch obstruction, even after successful surgical repair. The prevalence of systemic hypertension varies widely, from 20 to 68% [4–7]. This variability may be influenced by several factors, such as the definition of hypertension, methods of measuring blood pressure, timing of the repair, type of intervention and follow-up duration after arch repair [5, 8, 9].

Many theories have been advanced to explain the occurrence of this complication, including structural and functional abnormalities in the preaortic vessels, abnormalities in physiological mechanisms such as an imbalance within the autonomic nervous system, surgical stimulation of sympathetic nerve fibres causing a release of norepinephrine, altered responsiveness of the renin-angiotensin system, impaired vascular function and altered baroreceptor dysfunction [7, 9–11].

Recently, an abnormally shaped arch has been proposed as an important risk factor for the development of late systemic hypertension [12, 13]. Ou et al. [6] first categorized the shape of the aortic arch after coarctation repair into three types: gothic, crenel and roman. They reported that the gothic arch is associated with systemic hypertension, and requires harmonious repair of the arch obstruction to prevent or delay this condition in adulthood [6]. However, there have been few studies into the relationship between the surgical technique used for arch repair and the resulting arch geometry. Hence, in our present study, we evaluated the efficacy of a modified surgical technique in preventing a gothic arch.

MATERIALS AND METHODS

We performed a retrospective study of patients with arch obstruction who underwent an arch repair procedure by a single surgeon...
between January 2006 and August 2012 at Asan Medical Center and Konkuk University Medical Center, Seoul, Korea. The study was approved by the Institutional Review Boards at Asan Medical Center (study approval number: 2010-0465) and Konkuk University Medical Center (study approval number: KUH 1080019). The requirement for patient informed consent was waived.

All patients had an arch obstruction, combined with arch hypoplasia and an intracardiac anomaly. Arch hypoplasia was defined by using previously reported data [14]. Only those patients who underwent surgery prior to 6 months of age and had a biventricular physiology were included. A focused review of the medical and surgical records was undertaken and patient demographic, operative, echocardiographic, computed tomography (CT), cardiac catheterization, medication, perioperative and follow-up data were collected. Residual arch stenosis was defined as the presence of a resting peak pressure exceeding 20 mmHg across the repaired area. Early and late death was defined as mortality within and after 30 days following an arch reconstruction operation, respectively.

Surgical procedure

The surgical approach used in our patient cohort was a median sternotomy. A cardiopulmonary bypass (CPB) was established by interposing a 3.5-mm Gore-Tex vessel graft into the innominate artery at the aortic cannulation site, and venous drainage was accomplished via bicaval cannulation. After commencing CPB, patients were cooled to a state of moderate hypothermia (27–29°C). The operation was then performed using selective antegrade cerebral perfusion to protect the brain. Before 2008, we performed arch repair with conventional end-to-side anastomosis (ESA) or extended end-to-end anastomosis (EEEA), depending on the arch anatomy. However, after Ou et al.’s report in 2008 [6], we modified the arch repair technique. This modified technique was then performed as follows: on a beating heart, a pulmonary autologous patch was harvested from the anterior wall of the main pulmonary artery between the supracommissural level and the ductus arteriosus during cooling. The resulting defect to the main pulmonary artery was repaired with smaller-sized fresh autologous pericardium to reduce dilatation. After cardioplegia infusion, using selective cerebral perfusion, all of the ductal tissue was removed. An incision was made on the lesser curvature of the aortic arch, and the area of the defect was augmented with the harvested pulmonary autologous patch. A new longitudinal incision was made on the autologous pulmonary patch, and the descending aorta was anastomosed to that stoma with a 7/0 or 6/0 Prolene running suture (Ethicon, Somerville, NJ, USA) (Fig. 1).

Imaging work-up

A preoperative CT evaluation was performed in 43 infants [13 patients who underwent arch repair with a modified technique (modified group) and 30 patients who underwent arch repair with a conventional technique (conventional group)]. Seven patients could not undergo CT due to poor general health. Postoperatively, 31 patients (12 of the modified group and 19 of the conventional group), with parental consent, underwent a postarch repair CT evaluation. After arch reconstruction, the aortic arch shape was classified as gothic, crenel or roman based on the global geometry of the aortic arch, as previously reported [6]. Briefly, a gothic arch has a triangular form, a crenel arch has a rectangular form and a roman arch has a semicircular form, as presented in Fig. 2. Quantitative measurements were made of the transverse width (W), defined as the maximal horizontal distance between the midpoints of the ascending and descending aorta, and the height (H), defined as the maximal vertical distance between the W and the highest mid-point of the aortic arch, to calculate the height/width (H/W) ratio (Fig. 2).

Statistical analysis

The data were analysed using the Statistical Package for Social Science software (SPSS release 14.0 for Windows; SPSS, Chicago, IL, USA). Categorical variables are expressed as number and/or proportion. Continuous data are presented as median with interquartile range or mean ± standard deviation. Univariate comparisons of continuous variables were performed using the Student’s t-test or the Mann–Whitney U-test, as appropriate. Categorical variables were compared using the χ² test or Fisher’s exact test. For comparisons of more than two groups, an analysis of variance test was performed. In all comparisons, variables with a P-value of less than 0.05 were considered statistically significant.
There were no significant differences between the modified and conventional groups regarding clinical variables except for the selective cerebral perfusion time and the body weight at the time of operation (Table 3). There was one non-cardiac early death in the modified group, giving an early mortality rate of 2% (1/50). That patient died of massive haemorrhage after peritoneal dialysis catheter insertion as a complication on postoperative day 1. No patient showed systemic hypertension at the time of discharge. After discharge, all patients were regularly followed up by a cardiologist. The mean follow-up duration was 55.3 ± 26.7 months. At the last follow-up echocardiography, 9 patients showed a pressure gradient (mean, 30.9 ± 8.6 mmHg) at the arch repair site (1 in the modified group and 9 in the conventional group; P = 0.110). Among these cases, 3 patients in the conventional group underwent balloon coarctoplasty by catheter intervention. Reoperation for an arch reconstruction for obstruction relief of the left ventricular outflow tract was performed in 1 patient in the conventional group at 33 months after the initial arch repair. During follow-up, no significant pulmonary regurgitation, pulmonary artery aneurysm or stenosis of surgical lesion was detected in the modified group. One patient with IAA with VSD combined with subaortic stenosis died 84 days after arch repair and pulmonary artery banding due to a progressive left ventricular outflow tract obstruction and heart failure. This produced a late mortality rate of 2% (1/49). None of our patients showed significant systemic hypertension or needed cardiovascular medication during follow-up.

**Aortic arch geometry**

After arch repair, in a qualitative evaluation of aortic arch shape, we found that 9 patients of the conventional group showed abnormal gothic arch geometry (Table 4). In contrast, there were no patients with gothic arch geometry in the modified group (P = 0.005). However, crenel-type arch geometry was observed, with similar numbers in both groups. The H/W ratio was 0.79 ± 0.04 in gothic-type patients, 0.56 ± 0.06 in crenel-type patients and 0.63 ± 0.04 in roman-type patients. The aortic arch angles were 67.4 ± 7.1° in gothic-type, 82.2 ± 7.8° in crenel-type and 75.7 ± 7.0° in roman-type cases. These results indicate that the gothic arch geometry presented the greatest H/W ratio and narrowest arch angle.
A widely patent arch and a low incidence of restenosis are the principal points for successful arch repair. Introduction of a selective cerebral perfusion and tissue-to-tissue anastomosis technique has made it feasible to achieve the above goal [15, 16]. However, even after successful arch repair, there have been concerns about late systemic hypertension and related cardiovascular risks. Recently, mounting evidence has shown the relevance of the aortic arch geometry in hypertension occurrence [6, 13]. Ou et al. [6] stated that a proper H/W ratio of the aortic arch should be maintained after repair of arch obstruction, reporting that a gothic arch with high H/W is associated with abnormal intima-media thickness, higher aortic stiffness index, increased left ventricular mass index and increased systolic wave reflection, which explains the correlation between gothic arch geometry and late systemic hypertension [6, 12, 13, 17].

Insufficient attention has been paid to the relationship between the type of arch repair and resulting arch geometry, and few previous studies have compared different arch repair techniques in

### Table 2: Preoperative CT evaluation findings

<table>
<thead>
<tr>
<th>Variables</th>
<th>Modified group (n = 17)</th>
<th>Conventional group (n = 33)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Preoperative CT</td>
<td>13</td>
<td>30</td>
<td>0.23</td>
</tr>
<tr>
<td>Preoperative ascending aorta dimension (mm)</td>
<td>6.8 ± 1.2</td>
<td>7.3 ± 1.2</td>
<td>0.25</td>
</tr>
<tr>
<td>Preoperative ascending aorta Z-score</td>
<td>−3.2 ± 1.8</td>
<td>−2.5 ± 2.0</td>
<td>0.34</td>
</tr>
<tr>
<td>Preoperative proximal arch dimension (mm)</td>
<td>3.1 ± 0.6</td>
<td>3.5 ± 0.8</td>
<td>0.15</td>
</tr>
<tr>
<td>Preoperative proximal arch Z-score</td>
<td>−6.6 ± 2.0</td>
<td>−5.9 ± 1.7</td>
<td>0.27</td>
</tr>
<tr>
<td>Preoperative distal arch dimension (mm)</td>
<td>2.6 ± 0.6</td>
<td>2.8 ± 0.7</td>
<td>0.35</td>
</tr>
<tr>
<td>Preoperative distal arch Z-score</td>
<td>−6.4 ± 2.3</td>
<td>−5.8 ± 2.2</td>
<td>0.48</td>
</tr>
<tr>
<td>Preoperative descending aorta dimension (mm)</td>
<td>6.7 ± 0.7</td>
<td>6.6 ± 0.8</td>
<td>0.60</td>
</tr>
<tr>
<td>Preoperative descending aorta Z-score</td>
<td>0.7 ± 1.0</td>
<td>0.4 ± 1.1</td>
<td>0.53</td>
</tr>
<tr>
<td>Proximal arch/ascending aorta ratio</td>
<td>0.45 ± 0.09</td>
<td>0.47 ± 0.11</td>
<td>0.47</td>
</tr>
<tr>
<td>Distal arch/ascending aorta ratio</td>
<td>0.37 ± 0.08</td>
<td>0.37 ± 0.08</td>
<td>0.54</td>
</tr>
</tbody>
</table>

CT: computed tomography.

### Table 3: Baseline patient characteristics and clinical data

<table>
<thead>
<tr>
<th>Variables</th>
<th>Modified group (n = 17)</th>
<th>Conventional group (n = 33)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at operation</td>
<td>13.2 ± 12.9</td>
<td>23.9 ± 25.9</td>
<td>0.056</td>
</tr>
<tr>
<td>Weight at operation</td>
<td>3.0 ± 0.3</td>
<td>4.1 ± 2.0</td>
<td>0.005</td>
</tr>
<tr>
<td>Foetal echocardiographic diagnosis</td>
<td>11</td>
<td>17</td>
<td>0.22</td>
</tr>
<tr>
<td>Preoperative inotropes</td>
<td>8</td>
<td>13</td>
<td>0.77</td>
</tr>
<tr>
<td>Preoperative ventilator support</td>
<td>4</td>
<td>5</td>
<td>0.70</td>
</tr>
<tr>
<td>CPB time (min)</td>
<td>109.2 ± 25.6</td>
<td>98.6 ± 52.4</td>
<td>0.34</td>
</tr>
<tr>
<td>ACC time (min)</td>
<td>56.4 ± 18.4</td>
<td>48.6 ± 28.1</td>
<td>0.31</td>
</tr>
<tr>
<td>SCP time (min)</td>
<td>28.5 ± 6.2</td>
<td>17.1 ± 9.9</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Duration of postoperative ventilator support (days)</td>
<td>2.5 ± 0.9</td>
<td>2.9 ± 1.7</td>
<td>0.34</td>
</tr>
<tr>
<td>Duration of postoperative PICU stay (days)</td>
<td>6.0 ± 4.8</td>
<td>6.4 ± 4.2</td>
<td>0.80</td>
</tr>
<tr>
<td>Duration of hospital stay (days)</td>
<td>18.9 ± 10.4</td>
<td>17.7 ± 9.0</td>
<td>0.66</td>
</tr>
</tbody>
</table>

CPB: cardiopulmonary bypass; ACC: aortic cross-clamping; SCP: selective cerebral perfusion; PICU: paediatric intensive care unit; CT: computed tomography.

### Table 4: Postoperative quantitative and qualitative arch geometry

<table>
<thead>
<tr>
<th>Arch geometry</th>
<th>Width (mm)</th>
<th>Height (mm)</th>
<th>Angle (°)</th>
<th>Modified group (n = 12)</th>
<th>Conventional group (n = 19)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Roman</td>
<td>24.4 ± 7.5</td>
<td>15.5 ± 5.3</td>
<td>75.7 ± 7.0</td>
<td>8</td>
<td>7</td>
</tr>
<tr>
<td>Crenel</td>
<td>28.5 ± 6.0</td>
<td>16.0 ± 3.3</td>
<td>82.2 ± 7.8</td>
<td>4</td>
<td>3</td>
</tr>
<tr>
<td>Gothic</td>
<td>28.9 ± 7.7</td>
<td>22.8 ± 5.6</td>
<td>67.4 ± 7.1</td>
<td>0</td>
<td>9</td>
</tr>
</tbody>
</table>

Width: maximal horizontal distance between the mid-points of the ascending and descending aorta; height: maximal vertical distance between \( W \) and the highest mid-point of the aortic arch.

### DISCUSSION

A widely patent arch and a low incidence of restenosis are the principal points for successful arch repair. Introduction of a selective cerebral perfusion and tissue-to-tissue anastomosis technique has made it feasible to achieve the above goal [15, 16]. However, even after successful arch repair, there have been concerns about late systemic hypertension and related cardiovascular risks. Recently, mounting evidence has shown the relevance of the aortic arch geometry in hypertension occurrence [6, 13].
In terms of the resulting aortic arch geometry. Ou et al. [6] also did not note any relationship between the specific type of surgical technique and the resulting gothic arch geometry, but compared only end-to-end anastomosis (EEA) and sub-clavian arterial flap techniques.

In case of juxtaductal CoA or type A IAA, arch repair with simple resection and EEA could maintain normal arch geometry. However, for the type of CoA with arch hypoplasia or type B interruption of aortic arch, ESA or EEEA might distort the aortopulmonary space.

The ESA technique, introduced by Hanley and co-workers in 1996 [18], may cause narrowing between the ascending and descending aorta (W), as well as a decrease in height (H), as shown in Fig. 2A. Likewise, the EEEA technique introduced by Elliott et al. [19] in 1987 may result in a similar distortion of the aortopulmonary space and a tendency for a gothic arch to form. In 2002, a technical modification using an autologous pulmonary arterial patch for augmentation of the hypoplastic transverse arch was described [20]. The authors mainly focused on restenosis after arch repair, finding that this modified arch repair technique sufficiently augmented the hypoplastic arch without tissue deficit and foreign material use. They reported that this method resulted in a 100% actuarial freedom from recurrent arch obstruction with a median follow-up of 29 months. However, in terms of arch geometry, although the height of the arch was well maintained, a dilated main pulmonary artery remained. Moreover, the arch width was not effectively reduced because the descending aorta was anastomosed at the level of the isthmus. Consequently, this technique could result in a crenel-shaped arch geometry, as shown in Fig. 2B. We thus modified the technique with the aim of avoiding gothic arch geometry and sufficiently augmenting the hypoplastic aortic arch to appropriately reduce the width and maintain the height without using foreign material. To do this without tissue deficit, we used a pulmonic autograft, which has certain advantages such as reduced antigenicity, increased growth potential, which was a very important issue in infant patients, and a decreased risk of aneurysmal dilatation or calcification, as can occur when using a homograft or other prosthetic patch materials [21, 22]. Figure 3 shows the typical pre- and postoperative CT findings of infant patients with CoA with VSD who underwent our modified technique.

In our current study cohort, a gothic arch geometry was not observed in the modified group, unlike in the conventional group (0 vs 9; P = 0.005). However, a crenel-shaped arch geometry, which has been also associated with a higher incidence of systemic hypertension compared with the roman arch, was observed in 4 patients in the modified group and 3 in the conventional group [6]. Thus, for a more roman arch geometry, the arch repair technique should be refined to effectively reduce the width of the aortic arch and decrease the incidence of a crenel-type arch geometry.

In terms of restenosis of the aorta, residual arch stenosis resulting reintervention was detected only in 4 patients of the conventional group and in no patients of the modified group. As Roussin et al. reported, the use of a pulmonic autograft decreased restenosis and our results were also consistent with this [20]. Although this modified technique requires a slightly longer selective cerebral perfusion time (28.5 ± 6.2 min in the modified group vs 17.1 ± 9.9 min in the conventional group; P < 0.001), associated complications such as cerebral or renal ischaemic symptoms were not observed in our modified group of patients.

Systemic hypertension was not detected in either the modified or conventional groups at the time of discharge or during the follow-up period in this study. Hence, we found no difference in this complication between the two groups. The lack of hypertension could be partly explained as follows: firstly, our infant patients underwent repair in early infancy (mean age at the time of operation, 22.2 ± 26.5 days), an age that has previously been reported to have a significantly lower prevalence of hypertension [4, 23]. Secondly, we did not use a sub-clavian flap or prosthetic material for arch repair, material that has been associated with a higher prevalence of hypertension [4, 8, 24]. Thirdly, the mean follow-up duration of our study cohort was only 55.3 ± 26.7 months, which may be too short compared with previous studies to detect the development of systemic hypertension.

**Study limitations**

There are several limitations to this study of note. Firstly, due to its retrospective nature, the data are limited to the information available from medical records and the included patients showed several different baseline characteristics between the modified

![Figure 3: Pre- and postoperative CT findings for an infant with an aortic arch obstruction and ventricular defect in the modified group. (A) A dilated and enlarged main pulmonary artery (arrow) with varying degrees of hypoplasia of the transverse aortic arch. (B) Wide distance between the ascending aorta and descending aorta (W₁). (C) Maintaining the height (H₁ + H₂) and reducing the width (W₂) between the ascending and descending aorta by the modified technique can result in a roman arch.](image-url)
and conventional groups, including preoperative diagnosis and weight at the operation. Secondly, the follow-up duration was relatively short in terms of showing the effects on long-term cardiovascular morbidities such as systemic hypertension. Thirdly, the modified arch reconstruction technique has been used since 2008, which could be a selection bias by time period. Fourthly, a postoperative CT evaluation was not performed in all of our patients, which may also have contributed to a selection bias.

CONCLUSIONS

Surgical modification of aortic arch reconstruction with pulmonary autologous patch augmentation shows acceptable early outcomes with no restenosis and less physiological gothic arch geometry. This technique could subsequently be expected to reduce morbidity, including systemic hypertension. Despite the aforementioned limitations, our results suggest that our modified technique is a useful alternative option for arch repair in infants with arch obstruction combined with hypoplasia. For better long-term outcomes, constant circumspection and an effort to reconstruct the aortic arch to obtain a more physiological morphology with a harmonious H/W ratio are required.

Conflict of interest: none declared.

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