Aortic arch repair with extended end-to-side anastomosis in neonates and infants with transverse arch hypoplasia

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Abstract

Background/Aim: The use of patches to repair the aortic arch is believed to have a positive effect on long-term morbidity. In this study, perioperative and follow-up data of patients who underwent transverse arch repair with a patch were compared with the data of patients who underwent end-to-end anastomosis (ESA).

Methods: In this retrospective cohort study, the data of 27 patients (including 18 newborns) who underwent aortic arch repair at the Gazi Yasargil Education and Research Hospital between January 2018 and April 2023 were analyzed. The inclusion criteria included a diagnosis of proximal and distal transverse aortic arch hypoplasia, an age younger than 12 months of age, and the completion of aortic arch repair using cardiopulmonary bypass. Patients who underwent recoarctation repair due to residual obstruction, patients with single ventricular physiology, and patients who underwent aortic arch repair via a lateral thoracotomy without undergoing cardiopulmonary bypass were excluded from the study. The patients were divided into two groups. Group 1 included individuals who underwent aortic anterior wall expansion with autologous pericardium in addition to ESA; Group 2 included patients who underwent ESA only.

Results: The median age of the patients was 21 days (range: 6–365 days), and the median body weight of the cohort was 3.5 kilograms (range: 2.4–8.9 kilograms). Enlargement with autologous pericardial patch was applied to 11 patients (40.7%). Surgical procedures performed in addition to arch repair included eight ventricular septal defect closures, six instances of pulmonary banding, three atrial septal defect closures, and one subvalvular pulmonary stenosis repair. The in-hospital mortality rate was 11.1% (n=3). Those three patients died due to sepsis. The median follow-up period was 152 days (range: 10–1316 days). Recoarctation requiring re-intervention did not occur in any of the studied patients. The antegrade selective cerebral perfusion time was statistically significantly longer in patients who underwent aortic arch repair using a patch (P=0.03).

Conclusion: Repair of the arch with a patch may contribute to a reduction in long-term mortality and morbidity. However, there is a need for more comprehensive and long-term follow-up studies to verify these findings.

Keywords: aortic arch, aortic coarctation, congenital heart defects, infant, surgery
Introduction

Aortic arch hypoplasia is an anomaly that commonly requires surgical intervention during the neonatal period. The outcomes of aortic arch repair using the extended end-to-side anastomosis (ESA) technique are favorable [1]. Surgical techniques employed for the repair of transverse aortic arch hypoplasia include extended ESA, autologous pericardial patch, and bovine pericardial patch aortoplasty [2-5]. While the relatively shorter duration of surgery for direct anastomosis might be advantageous, it has been argued that recoarctation might occur due to changes in arch geometry [2]. On the other hand, patch aortoplasty preserves the geometry of the arch aorta and reduces tension at the anastomotic site [6,7]. In this study, we compare aortic arch repair results in patients with aortic arch hypoplasia.

Materials and methods

Ethical declaration

The study was conducted in accordance with the Declaration of Helsinki, and the study protocol was approved by the Diyarbakır Gazi Yasargil Training and Research Hospital Ethics Committee (Date: May 26, 2023, No: 508).

Definitions

The transverse aortic arch is the name given to the part of the aorta between the innominate artery and the left subclavian artery. Proximal transverse arch hypoplasia is indicated when the ratio of the outer diameter of the proximal arch to the diameter of the ascending aorta is less than 0.6, and distal transverse arch hypoplasia is defined as a ratio of the outer diameter of the distal arch to the ascending aorta diameter below 0.5 [6,8]. Z-scores of the aortic arch were calculated using transthoracic echocardiography [9]. In postoperative transthoracic echocardiography, recoarctation was diagnosed if a gradient exceeding 20 mmHg was measured at the surgically repaired area [10].

Patient population

A total of 27 patients diagnosed with transverse arch aortic hypoplasia and aortic coarctation who underwent aortic arch repair from January 2018 to April 2023 at the Gazi Yasargil Education and Research Hospital in Diyarbakır, Turkey were included in the study. Preoperative, intraoperative, and postoperative data were recorded using the hospital's database. Follow-up visits were planned at 1, 3, and 6 months after discharge, every 6 months for the following 18 months, and subsequently annually. Follow-up data were collected by reviewing echocardiography reports from the outpatient clinic system.

The inclusion criteria included a diagnosis of proximal and distal transverse arch aortic hypoplasia, an age younger than 12 months, and completion of aortic arch repair with cardiopulmonary bypass via median sternotomy. Patients undergoing re-coarctation repair due to residual obstruction, those with single ventricle physiology, and those undergoing aortic arch repair via lateral thoracotomy without cardiopulmonary bypass were excluded. Seven of the patients were on preoperative mechanical ventilator support. Milrinone was given to eight patients, and adrenaline treatment was given to three patients. Ten of the patients had undergone preoperative balloon angioplasty. The patients were divided into two groups:

Group 1 included individuals who underwent aortic anterior wall expansion with autologous pericardium in addition to ESA;

Group 2 included individuals who underwent ESA only. The patients' ages, genders, body weights, diagnoses, accompanying anomalies, antegrade selective cerebral perfusion (ASCP) times, cross-clamp times, cardiopulmonary bypass times, postoperative complications, vasoactive inotrope scores, mechanical ventilation times, length of stay in the intensive care unit, length of hospital stay, follow-up times, and preoperative transverse arch Z-scores were compared.

Surgical technique

Following median sternotomy, all of the patients underwent a cardiopulmonary bypass via the innominate artery and bicaval venous cannulation. Antegrade selective cerebral perfusion and systemic hypothermia at 28°C were employed. The transverse aortic arch, innominate artery, left carotid artery, left subclavian artery, isthmus, and descending aorta were individually dissected. After clamping the innominate artery, aortic arch branches, and descending aorta, a single dose of 20 ml/kg del Nido cardioplegia solution was administered from the aortic root. The isthmus and proximal descending aorta were divided, and ductus arteriosus tissue on the side of the descending aorta was completely removed. A longitudinal incision was made on the lesser curvature of the aortic arch between the innominate artery and the isthmus. The descending aorta was sutured to the posterior wall of the aortic arch using a continuous suturing technique. In cases where mobilization of the descending aorta was limited and there might be tension at the anastomotic site, the lesser curvature of the transverse arch was widened using an autologous pericardial patch treated with glutaraldehyde. In patients with adequately mobilized descending aorta, the anterior wall of the aorta was sutured to the native aorta using the same suture, completing the anastomosis. After deairing and removing the clamps, full-body perfusion was resumed.

Statistical analysis

Statistical analyses were performed using SPSS statistical software version 23. The Kolmogorov-Smirnov test was used to assess normality. Descriptive statistics were provided as median (minimum-maximum) for continuous variables and as percentages for categorical variables. The Mann-Whitney U test was used for binary comparisons of continuous variables between independent groups, and the chi-squared test was used for categorical variables. P-values less than 0.05 were considered to be statistically significant.

Results

Patient characteristics

Between January 2018 and April 2023, a total of 27 consecutive patients underwent aortic arch repair surgery at the Gazi Yasargil Education and Research Hospital. Patient information was retrospectively obtained from the hospital's database. Of the 27 patients, 51.8% (14) were male. Eighteen patients (63%) were newborns, and five (18.5%) were premature. The median age of the patients on the day of surgery was 21 days (range: 6–365 days), and the patients’ median body weight was...
3.5 kilograms (range: 2.4–8.9 kilograms). The demographic characteristics of the patients are listed in Table 1. Sixteen patients (59.2%) were intubated preoperatively. Eleven patients (40.7%) had identified genetic anomalies. The most common genetic anomaly was Down syndrome (n=4, 36.4%), followed by Turner syndrome (n=1, 9.1%) and DiGeorge syndrome (n=1, 9.1%). Other anomalies included genitourinary anomalies, limb anomalies, orofacial anomalies, anal atresia, and a dysmorphic facial appearance (n=5, 45.4%). A bicuspid aortic valve was present in 10 patients (37.1%), persistent left superior vena cava was present in three patients (11.1%), and aberrant right subclavian artery was present in two patients (7.4%).

**Intraoperative and postoperative outcomes**

Sixteen patients (59.3%) underwent direct ESA, and 11 patients (40.7%) underwent ESA with expansion of the anterior wall using an autologous pericardial patch. Additional cardiac surgical procedures other than patent ductus arteriosus ligations and division were performed in 15 patients (55.6%). Those included eight (29.6%) ventricular septal defect closures (using an autologous pericardial patch), six (22.2%) pulmonary bandings, three (11.1%) atrial septal defect closures, and one (3.7%) pulmonary valvotomy (Figure 1). The mean (standard deviation) duration of ASCP was 35.6 (13.3) minutes; the mean (standard deviation) aortic cross-clamp time was 59.2 (26.9) minutes. The mean (standard deviation) cardiopulmonary bypass time was 109.4 (38.1) minutes. Antegrade selective cerebral perfusion time was statistically longer in patients who were repaired using a patch compared with patients in the other group (P=0.03).

**Postoperative hospital mortality**

Postoperative hospital mortality was recorded for three patients (11.1%). Two patients succumbed to sepsis during the early postoperative period (0–30 days), and one patient who understood a prolonged intensive care unit stay due to fungal sepsis passed away in the late postoperative period (after one month). Sepsis was the most common complication and was observed in seven patients (25.9%). Four of those patients were successfully treated with appropriate antibiotic therapy. Other complications included acute renal failure requiring peritoneal dialysis (n=3, 11.1%), arrhythmia (n=2, 7.4%), acute respiratory distress syndrome (n=1, 3.7%), pneumonia (n=1, 3.7%), and chylothorax (n=1, 3.7%). The postoperative complications are shown in Figure 2. The median follow-up duration for all of the patients was 152 days (range: 10–1316 days). No recoarctation was detected on echocardiography at discharge or during follow-up, and no interventions were required. The patients’ perioperative data are listed in Table 2.

![Figure 1: Additional cardiac surgical procedures performed on patients](image)

**Figure 2: Postoperative complications**

ASD: atrial septal defect, VSD: ventricular septal defect

### Table 1: Demographic and preoperative data

<table>
<thead>
<tr>
<th>Patient characteristics</th>
<th>ESA + patch (n=11)</th>
<th>ESA (n=16)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender, n, (%)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>5 (45.5%)</td>
<td>8 (50%)</td>
<td>0.81**</td>
</tr>
<tr>
<td>Male</td>
<td>6 (54.5%)</td>
<td>8 (50%)</td>
<td></td>
</tr>
<tr>
<td>Age at surgery (days), median (min-max)</td>
<td>24 (10-365)</td>
<td>21 (6-120)</td>
<td>0.31*</td>
</tr>
<tr>
<td>Weight at surgery (kg), median (min-max)</td>
<td>4 (2.86-8.90)</td>
<td>2.87 (2.41-7.78)</td>
<td>0.07**</td>
</tr>
<tr>
<td>Neonates n, (%)</td>
<td>6 (54.5%)</td>
<td>12 (75%)</td>
<td></td>
</tr>
<tr>
<td>Preoperative LVEF (%), median (min-max)</td>
<td>65 (40-78)</td>
<td>65 (40-74)</td>
<td>0.48*</td>
</tr>
<tr>
<td>Transverse aortic arch Z score, median (min-max)</td>
<td>-4.7 [-9.1-3.6]</td>
<td>-5.1 [-8.2-3.6]</td>
<td>0.44*</td>
</tr>
</tbody>
</table>

**ESA:** End-to-side anastomosis, **LVEF:** Left ventricular ejection fraction, min-max: minimum-maximum; *: Mann-Whitney U test; #: Chi-squared test

### Table 2: Perioperative findings of the patients

<table>
<thead>
<tr>
<th>Parameters</th>
<th>ESA + patch (n=11)</th>
<th>ESA (n=16)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>CPB time (min), median (min-max)</td>
<td>11 (6.2-192)</td>
<td>10 (4.0-152)</td>
<td>0.34*</td>
</tr>
<tr>
<td>ACC time (min), median (min-max)</td>
<td>53 (10-112)</td>
<td>46 (18-105)</td>
<td>0.55*</td>
</tr>
<tr>
<td>ASCP time (min), median (min-max)</td>
<td>37 (26-65)</td>
<td>38 (16-70)</td>
<td>0.03*</td>
</tr>
<tr>
<td>Postoperative gradient (mmHg), median (min-max)</td>
<td>5 (0-28)</td>
<td>6 (3-39)</td>
<td>0.71*</td>
</tr>
<tr>
<td>ICU stay (days), median (min-max)</td>
<td>11 (5-40)</td>
<td>14 (5-64)</td>
<td>0.21*</td>
</tr>
<tr>
<td>Mechanical support time (h), median (min-max)</td>
<td>5 (1-12)</td>
<td>5 (2-26)</td>
<td>0.24*</td>
</tr>
<tr>
<td>In-hospital time (days), median (min-max)</td>
<td>17 (9-86)</td>
<td>18 (7-70)</td>
<td>0.74*</td>
</tr>
<tr>
<td>Vascular isotropic score, median (min-max)</td>
<td>12 (7-27)</td>
<td>11 (7-20)</td>
<td>0.43*</td>
</tr>
<tr>
<td>In-hospital mortality (0-30 days) n</td>
<td>1</td>
<td>1</td>
<td>0.78*</td>
</tr>
<tr>
<td>Follow-up time (days), median (min-max)</td>
<td>167.5 (10-1164)</td>
<td>123 (17-1116)</td>
<td>0.69</td>
</tr>
</tbody>
</table>

**CPB:** Cardiopulmonary bypass, min-max: minimum-maximum, *: Mann-Whitney U test; #: Chi-squared test
Discussion

The goal of surgically treating patients with aortic coarctation in conjunction with arch hypoplasia is to achieve the least number of repeat interventions and ensure adequate arch growth over the long term [11-13]. To this end, the arch geometry should be restored as close to normal as possible. Surgical repair with extended ESA in the segment between the innominate artery and the left common carotid artery, where proximal arch hypoplasia is present, can be relatively challenging [12-14]. Researchers have differing viewpoints as to the most appropriate repair strategy for this part of the hypoplastic arch. Some centers prefer repairing the arch with median sternotomy and a cardiopulmonary bypass; others opt for a posterolateral thoracotomy without stopping the heart [11,15,16]. However, aortic repair using cardiopulmonary bypass necessitates consideration of its potential harmful effects. In our study, aortic arch repair was performed in all patients after median sternotomy with cardiopulmonary bypass and ASCP.

The use of transverse aortic arch repair using ESA is debated in the literature. Some studies have demonstrated favorable and positive results with ESA in cases of proximal transverse arch hypoplasia [16,17]. However, satisfactory results could not be obtained with ESA in some patients with a large anastomotic area [2,18]. Reducing tension at the anastomotic site is crucial for preventing postoperative recoarctation and is the most significant factor in the treatment of transverse arch hypoplasia [18,19]. In patients with pronounced distance between the hypoplastic segment of the transverse arch and the descending aorta, creating an anastomosis without tension may not always be possible [3]. Additionally, tension at the anastomotic site can lead to late recoarctation [3,18]. Reducing anastomotic tension and creating a smooth arch geometry can be achieved with patch aortoplasty. The use of a patch can sufficiently expand the hypoplastic segment to prevent recoarctation and achieve a relatively normal geometry [18,20]. In a study comparing ESA and patch aortoplasty techniques, a lower incidence of recoarctation was reported in the patch group [21]. Li et al [21] reported that the ESA method was preferable to patch aortoplasty in patients for whom ESA was suitable (i.e., individuals in which descending aorta mobilization was not limited and aortic arch geometry was normal). The authors noted that the ESA method was associated with fewer procedures and a shorter ASCP time. Among our patients, 11 (40.7%) had a relatively long distance between the narrow segment and the descending aorta (Group 1). Therefore, we enlarged the lesser curvature of the aorta using an autologous pericardial patch to reduce tension at the anastomotic site. Antegrade selective cerebral perfusion times were statistically significantly longer in those patients than in Group 2 patients.

The relationship between transverse arch hypoplasia and intracardiac defects has been described by Rudolph et al [22]. Those authors found that intracardiac defects may lead to decreased blood flow in the aortic arch, resulting in underdevelopment. Furthermore, Conte et al [23] reported that one-stage repair yielded better outcomes than two-stage repair in 307 newborns with aortic coarctation and accompanying complex cardiac defects. The feasibility and superiority of simultaneous repair of intracardiac defects with arch repair have been reported by other studies as well [24,25]. In our investigation, 70.4% of patients with transverse aortic arch hypoplasia had one or more accompanying intracardiac defects. At our center, we perform simultaneous (one-stage) repair procedures for patients with aortic coarctation/hypoplasia associated with perimembranous ventricular septal defect (VSD). For patients with multiple VSDs, midmuscular VSD, or atrioventricular septal defects, we prefer the arch repair and pulmonary banding (two-stage) procedure.

Limitations

The retrospective nature of our study, its small sample size, and its limited follow-up duration are its most significant limitations. Furthermore, our investigation was a single-center study. Therefore, increasing the number of cases, supporting the study’s findings with prospective comparative research, and investigating long-term outcomes with extended follow-up are necessary.

Conclusions

Repairing the aortic arch in newborns and infants with transverse arch hypoplasia using extended ESA is a safe, effective, and promising procedure with low rates of early reintervention and favorable short-term outcomes. It allows for the repair of accompanying intracardiac defects and preserves the growth potential of the aortic arch via tissue-on-tissue reconstruction. Aortic arch repair can be done with or without a patch. However, the use of a patch on the anterior aortic wall of the aorta in aortic arch repair reduces tension on the anastomotic line, protects the growth of the aortic arch, and helps prevent recoarctation.

References