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Management of aortic arch hypoplasia in neonates and infants

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Abstract

Objectives: Surgical management of aortic arch hypoplasia (AAH) with associated intracardiac anomalies is a challenge in newborns. We reviewed the characteristics and outcomes of neonates and infants who underwent pulmonary artery banding concomitant to arch repair and single-stage total repair at our institution.

Methods: Medical records of 60 patients undergoing aortic arch reconstruction for AAH from 2014 to 2019 were retrospectively reviewed. Twenty-five patients were female (41.6%), and the age of the patients ranged from 4 to 120 days (median, 19.5 days). The patients were divided into two groups: Group 1 (23 patients) underwent pulmonary artery banding concomitant to arch repair, and Group 2 (37 patients) underwent single-stage total repair in addition to arch repair. All arch repair procedures consisted of an extended (to the midportion of the ascending aorta) patch aortoplasty.

Results: Postoperative early mortality occurred in 12 patients, eight in Group 1 (34.8%) and four in Group 2 (10.8%). There was an early survival advantage in Group 2 ($p = .019$). Recoarctation occurred in 13 cases (21.6%), and 11 (18.3%) of them required reintervention (balloon angioplasty: 7, reoperation: 4). On univariate analysis, risk factors associated with death were pulmonary artery banding (hazard ratio [HR], 0.44; confidence interval [CI], 0.09–2; $p = .019$), prematurity (HR, 4.67; CI, 1.34–16.18; $p = <.001$), preoperative mechanical ventilation support requirement (HR, 0.048; CI, 0.52–6.39; $p = .048$), and functional single ventricle (HR, 0.43; CI, 0.1–1.86; $p = .006$). The mean duration of follow-up was 21.9 ± 15.1 months, and there was no late death in either group.

Conclusion: Single-stage repair of AAH with intracardiac pathologies has better results than palliation, according to survival rates and postoperative results. The use of the patch augmentation technique in AAH is valid and associated with an acceptable incidence of recurrent arch obstruction.

KEYWORDS

aortic arch plasty, congenital heart disease, hypoplastic aortic arch

1 | INTRODUCTION

Aortic arch hypoplasia (AAH) involving all segments of the arch usually occurs in neonate and infants with intracardiac anomalies.[1–3] Discrete aortic coarctation is rare in the infantile period, and up to 31% of these patients have an associated degree of AAH.[1] Although there are still no definite diagnostic criteria, AAH is generally considered present when the aortic arch diameter between the

innominate artery and the left common carotid artery is <60%, between the left common carotid artery and the left subclavian artery is <50%, and the aortic isthmus is <40% of the ascending aorta diameter.[1] Surgical management of AAH is a surgical challenge. Associated intracardiac pathologies are present with varying severity ranging from a simple ventricular septal defect to hypoplastic left heart syndrome, which may affect decision making and outcomes. There is also great diversity in the myocardial surgical repair techniques and cerebral protection strategies. Moreover, postoperative recurrent aortic arch obstruction remains a significant problem mid-to-long term.[3,4]

Decision making between single stage repair and a staged approach is not only dependent on the intracardiac anomaly but also the patients' general condition and the surgeons' preference. In the current study, we reviewed our early- and midterm results in neonates and infants with AAH and intracardiac anomalies in terms of single stage or staged repair.

2 | MATERIALS AND METHODS

Medical records of 60 consecutive patients who underwent aortic arch reconstruction between January 2014 and December 2019 were retrospectively reviewed. Patients with hypoplastic left heart syndrome, interrupted aortic arch, and isolated coarctation were excluded from the study. All but three patients had associated intracardiac anomaly. Our study received ATADEK-2020/2 (numbered 2020-2/6) Ethics Committee approval on 11/02/2020. We defined AAH according to the described criteria.[1] Transthoracic echocardiography was used for the diagnosis of all patients. Their aortic arch anatomy was examined in detail. Computerized tomography angiography was performed upon any suspicion on the echocardiographic diagnosis. The mean diameter of the proximal transverse arch (between the innominate and the left carotid arteries) was 3.61 ± 0.9 mm (the mean *Z* score: −5.21 ± −1.44), the distal transverse arch (between the left carotid and the left subclavian arteries) was 3.6 ± 0.91 mm (the mean *Z* score: −3.24 ± −1.38), and the isthmic arch (between the left subclavian and the ductus arteriosus) was 2.98 ± 0.93 mm (the mean *Z* score: −3.98 ± −1.65).

The age of the patients ranged from 4 to 120 days (median, 19.5 days), their weight ranged from 2 to 5.3 kg (mean, 3.2 ± 0.73 kg), and their body surface area (BSA) ranged from 0.16 to 0.28 m^2 (mean, 0.21 ± 0.02 m^2). Forty-six patients (76.6%) were neonates, and 11 patients were premature (18.3%). The male-to-female ratio was 35/25 (58.4%/41.6%).

The patients were divided into two groups. Group 1 consisted of 23 patients who underwent palliation with pulmonary artery banding concomitant to aortic arch repair. Group 2 consisted of 37 patients who underwent single-stage total repair. The main characteristics, associated intracardiac anomalies, operative details, and perfusion strategies for both groups are described in Tables 1 and 2.

Ductal dependency and prostaglandin E1 infusion were necessary in 45 patients (75%), and 42 patients (70%) had congestive heart failure preoperatively. Sixteen patients (26.6%) required mechanical ventilatory support before the operation. Three patients had trisomy 21 and 3 had Williams syndrome. Twelve patients (20%) had poor preoperative clinical conditions. Five of the patients were treated for sepsis, four had congenital pneumonia, two had acute renal failure, and the last one had multiorgan failure. Three patients had undergone surgery due to esophageal atresia ($n = 2$) and anal atresia ($n = 1$) before cardiac operations.

TABLE 1 Associated cardiac anomalies are listed

Diagnosis	Total ($n = 60$)	Palliation ($n = 23$)	Total correction ($n = 37$)
VSD	22 (36.7%)	6	16
Multiple/muscular VSD	6 (10%)	4	2
Double outlet right ventricle with subpulmonic VSD	8 (13.3%)	2	6
TGA, VSD	6 (10%)	0	6
Bicuspit aortic valve	5 (8.3%)	2	3
Tricuspid atresia	4 (6.7%)	4	0
isolated aortic arch hypoplasia	3 (5%)	0	3
Balanced CAVSD	2 (3.3%)	2	0
Double inlet left ventricle	2 (3.3%)	2	0
AP window	1 (1.7%)	0	1
Unbalanced CAVSD	1 (1.7%)	1	0

Abbreviations: AP, aortopulmonary; CAVSD, complete atrioventricular septal defect; TGA, transposition of the great arteries; VSD, ventricular septal defect.

TABLE 2 Perioperative characteristics and perfusion strategy of the study

Characteristics	Total ($n = 60$)	Palliation ($n = 23$)	Total correction ($n = 37$)	p value
Male gender, *n* (%)	35 (58.3%)	9 (39.1%)	26 (70.2%)	**.017**
Weight at surgery, kg	3.22 ± 0.74	3.01 ± 0.78	3.35 ± 0.69	.084
BSA at surgery, m^2	0.21 ± 0.03	0.20 ± 0.03	0.22 ± 0.03	**.044**
Mean age at surgery, days	25.70 ± 22.65	23.65 ± 24.21	26.97 ± 21.87	.280
Weight ≤2.5 kg, *n* (%)	11 (18.3%)	7 (30.4%)	4 (10.8%)	.056
Genetic syndromes	6 (10%)	2 (8.6%)	4 (10.8%)	.62
Preoperative mechanical ventilatory support	16 (26.67%)	7 (30.4%)	9 (24.3%)	.6
Proximal transverse arch *Z* score, median range	−5.38 (−8.20 to −2.20)	−5.58 (−8.20 to −2.20)	−5.25 (−7.65 to −2.40)	.083
Distal transverse arch *Z* score, median range	−3.26 (−6 to −0.4)	−3.65 (−6 to −0.4)	−3.2 (−6 to −0.4)	**.045**
Isthmic arch *Z* score, median range	−3.82 (−6.79 to −1.32)	−4.56 (−6.23 to −1.32)	−3.8 (−79 to −1.17)	.226
Operative parameters				
CPB time, min, mean ± *SD*	88.9 ± 47.3	56.9 ± 10.4	108.7 ± 50.5	**<.001**
Cross-clamp time, min, mean ± *SD*	38.4 ± 35	12.6 ± 6.5	54.4 ± 36	**<.001**
ACP time, min, mean ± *SD*	22.4 ± 7.4	21.5 ± 6.6	23 ± 7.8	.709

Note: Bold values are statistically significant.
Abbreviations: ACP, antegrade cerebral perfusion; BSA, body surface area; CPB, cardiopulmonary bypass.

3 | SURGICAL REPAIR TECHNIQUE

Median sternotomy and cardiopulmonary bypass (CPB) was performed on all patients. The thymus was totally excised and the pericardium was harvested. A straight tip 8Fr (2.7 mm) arterial cannula (RMI, Edwards Lifesciences LLC) was inserted into the distal ascending aorta and advanced into the innominate artery for antegrade selective cerebral perfusion (ASCP) during the aortic arch repair. No additional ductal aortic cannulation was performed. Distal perfusion was maintained through the arterial duct by snaring both pulmonary artery branches and keeping the heart filled during cooling. Near-infrared spectroscopy (NIRS) monitoring was performed. Usually, it took approximately 10–15 min to reach the desired core temperature. The Alfa-stat strategy was used in acid–base management for cerebral protection. Aortic arch repair was performed first in all patients. Aortic arch vessels, duct and descending aorta were dissected free. During moderate hypothermia (26°C), an aortic clamp was inserted between the innominate and left carotid artery to maintain selective cerebral and coronary flow. The flow rate was adjusted to approximately 50 ml/kg per min. The mean blood pressure of the right radial artery was sustained at approximately 40–50 mmHg, and the mean hematocrit level was sustained at approximately 30%. Yasargil neurovascular clips were used to close the left carotid and left subclavian arteries. The descending aorta was clamped as far distally as possible with a side clamp. The arterial duct was ligated and resected from the aortic end. The aortic arch was incised until proximal aortic clamp. After all ductal tissue was resected, the distal descending aorta was anastomosed to the aortic arch to form a native posterior wall. The distal descending aorta was incised anteriorly and a pericardial patch was sutured to augment the anterior wall of the aortic arch. When the suture line approaches to the aortic clamp, the innominate artery was snared over the aortic cannula and ASCP commenced. At the same time, tepid blood cardioplegia was given. The aortic clamp was moved to the proximal ascending aorta and the arch incision was extended to the midportion of the ascending aorta. The remainder of the pericardial patch was sutured to cover the rest of the aortic incision. After deairing of the aorta, clamps were removed, the aortic cannula moved into the aortic arch and whole body perfusion was begun. During rewarming, pulmonary artery banding or intracardiac repairs were performed.

3.1 | Data collection and availability

Data collection was performed by review from our institution. The baseline characteristics, perioperative and postoperative results were documented. In addition to early and late mortality, postoperative early and late morbidities were recorded. Microsoft Excel (Microsoft Corp.) was used to extract data. The data that support the findings of this study are available from the corresponding author, [author initials], upon reasonable request.

3.2 | Statistical analysis

Statistical analyses were performed by a biostatistician using SPSS Statistics version 21.0 (IBM). The normality of data was analyzed using Kolmogorov-Smirnov. The descriptive statistics were reported as the mean ± *SD*, medians, and ranges (min-max). The independent sample *T* test and the Mann–Whitney *U* test were used to determine the average difference between the two independent groups for parametric and nonparametric variables, respectively. The relationship between two sets of data was analyzed by Spearman's rank correlation test. Overall survival analyses of the groups were evaluated with Kaplan–Meier curves, and differences were tested with a log-rank test. All comparative tests were two-tailed, and a *p* value of <.05 was considered to be statistically significant. Effects of covariates on the possibility of survivals in univariate analysis are reported as hazard ratio with the 95% confidence interval.

4 | RESULTS

Operative and postoperative results are listed in Table 2. The mean BSA of the patients in Group 1 was lower than in Group 2 ($p = .044$), and the number of premature patients was higher in Group 1 ($p = .056$). In Group 1, four patients underwent atrial septectomy, and two underwent aortic valve commissurotomy because of bicuspid aortic valve, additionally. Pulmonary artery banding was performed in one of these two patients who underwent aortic valve commissurotomy due to multiple VSD. In Group 2, 18 patients underwent VSD closure, 12 patients underwent arterial switch operation (ASO) with VSD closure, and one underwent aorto-pulmonary window repair (Figure 1). Although three of the remaining six patients had a bicuspid aortic valve, the valve did not need commissurotomy, and similar to the three patients with isolated arch hypoplasia, these patients underwent isolated aortic arch repair. The mean ASCP times were 22.4 ± 7.4 min. The mean CPB and aortic cross-clamping (CC) times were significantly longer in Group 2 (CPB time: 56.9 ± 10.4 min in Group 1 vs. 108.7 ± 50.5 min in Group 2, $p < .001$; CC time: 12.6 ± 6.5 min in Group 1 vs. 54.4 ± 36 min in Group 2, $p < .001$).

In Group 1, 10 patients underwent pulmonary artery banding despite having functional biventricle and VSD. Of these 10 patients, 4 (40%) were premature, 6 (60%) had low body weight (≤2.5 kg), and 3 (30%) had poor preoperative condition or required mechanical ventilation support. Only three patients did not have any comorbidities and VSDs closing was postponed to the second stage due to anatomical location or size. They had large subaortic, apical muscular and large inlet-outlet lying VSDs, respectively.

Hospital mortality was 34.8% and 10.8%, respectively (8/23 in Group 1 and 4/37 in Group 2, $p = .024$). Of the eight patients in Group 1, 6 (75%) were neonate, 4 (50%) were male, 4 (50%) had low body weight (<2.5 kg), 5 (62.5%) had functional single ventricle, and 5 (62.5%) had preoperative poor condition or required mechanical ventilation support. Bleeding complications occurred in two patients, low cardiac output occurred in three patients, and they died due to sepsis in the postoperative course. One patient had sudden cardiac arrest on postoperative day 2 and died despite resuscitation. Extracorporeal membrane oxygenation (ECMO) support was not used in this patient because of low body weight and poor preoperative condition. The remaining two patients died due to *Klebsiella pneumoniae* sepsis and Enterococcus faecium mediastinitis after the long postoperative course, respectively. Of the four patients in Group 2, all patients were neonate and male, 1 (25%) had low body weight (<2.5 kg), and 3 (75%) needed preoperative sepsis treatment or required mechanical ventilation support. One patient underwent reoperation due to mitral valve insufficiency after the perimembranous

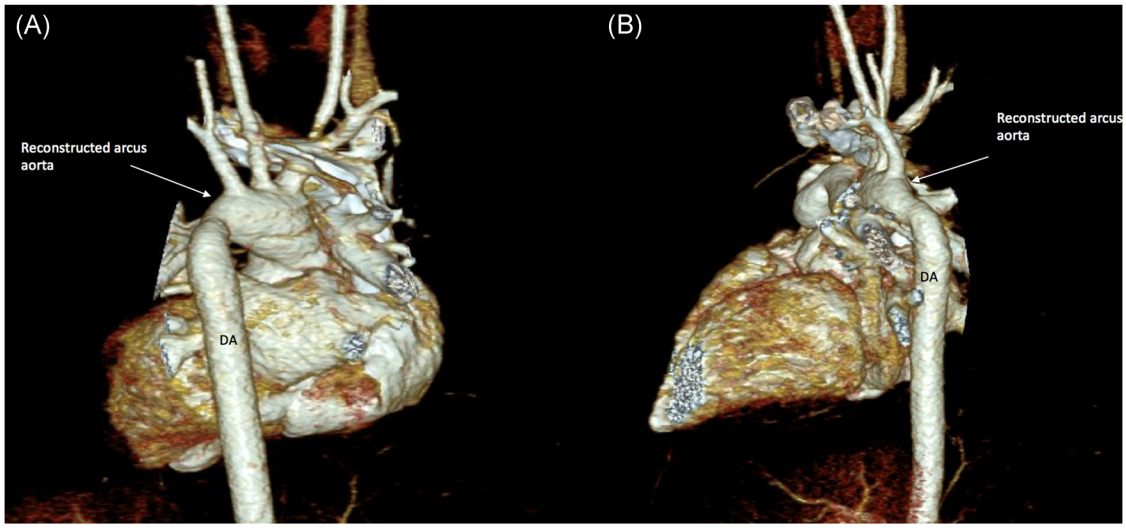

FIGURE 1 Postoperative CT scan at the 6th month with a 3-D reconstruction of a neonate who underwent arterial switch and aortic arch reconstruction operation because of Taussig Bing anomaly with AAH viewed laterally (A, B). AAH, aortic arch hypoplasia; CT, computed tomography; DA, descending aorta.

VSD closure; this patient and another patient died because of sepsis after long postoperative course. ECMO support was used in the remaining two patients due to low cardiac output. One of them weaned from ECMO but could not survive and died due to multiorgan failure and the other died on ECMO because of *K. pneumoniae* sepsis.

The median duration of intensive care unit (ICU) and hospital stay were 11 days (range, 2–228), and 19 days (range, 2–230), respectively. Although the median length of ICU stay was not statistically significant, it was longer in Group 1 (15 days to 10 days, $p = .45$). In addition, the median length of hospital stay was not different between two groups (19 days to 19 days, $p = .93$). Sternal closure was delayed in 35 cases (58.3%) by a mean of 4.3 ± 5.9 days (range, 1–33), and there was no statistically significant difference between the two groups (12/23 in Group 1 and 23/37 in Group 2, $p = .44$). However, the mean duration of delayed sternal closure time was significantly higher in Group 1 (6.8 ± 8.7 vs. 2.7 ± 2.1, $p = .044$). Twenty-three patients (39.6%) (10/23 vs. 13/37, $p = .48$) needed a long duration of mechanical ventilation (>7 days), and 24 patients needed prolonged ICU stay (>15 days) (12/23 vs. 11/37, $p = .12$). VAC therapy was performed due to mediastinitis or delayed sternal closure in five cases (8.3%) and was significantly higher in Group 1 (4/23 vs. 1/37, $p = .045$). Three patients (5%) needed re-exploration for bleeding, and two patients (3.3%) experienced supraventricular dysrhythmias. Eight patients (13.3%) showed postoperative low cardiac output syndrome and that was statistically significantly higher in Group 1 (6/23 vs. 2/37, $p = .022$). Sudden cardiac arrest occurred in four cases (6.6%), and all of them were in Group 1 ($p = .009$). Two patients needed postoperative ECMO support due to extracorporeal cardiopulmonary resuscitation or low cardiac output. Unfortunately, although one patient weaned from ECMO, both patients died due to sepsis. Tracheostomy was required due to the long duration of mechanical ventilation in four cases (6.6%). Sepsis occurred in seven cases (11.6%) (5/23 vs. 2/37, $p = .055$), and one of them had chylothorax. Two patients underwent left diaphragm plication because of left diaphragm paralysis in the early postoperative period. Peritoneal dialysis was required in three cases (5%) because of renal insufficiency and/or positive fluid balance. Minor neurological events occurred in three cases (5%). Three patients (5%) needed antihypertensive therapy while discharged from the hospital. Outcomes and complications were presented in Table 3.

Follow-up was completed for 93.75% (45/48) survivors with a mean duration of 21.9 ± 15.1 months (range, 2–58 months). Kaplan–Meier survival for both groups is shown in Figure 2, and there was an early survival advantage in Group 2 ($p = .019$). Risk factors affecting survival are listed in Table 4. No mortality was observed during follow-up.

Recoarctation occurred in 13 cases (27%) and 11 (22.9%) of them required reintervention (Figure 3). We used glutaraldehyde-treated autologous pericardium in 21 patients, bovine pericardium in 19 patients, and porcine pericardium in 20 patients, and there was no statistically significant relationship between patch types and recoarctation ($p = .77$). Moreover, there was no association between the *Z* scores of the proximal aortic arch, distal aortic arch, transverse aortic arch, and recoarctation. Similarly, the incidence of reintervention was not statistically significant between the two groups ($p = 0.89$; Figure 4). Among 13 patients, seven patients underwent balloon dilation angioplasty, four patients required reoperation as patch aortoplasty after unsuccessful balloon dilatation, and the last two patients were closely followed up by medical treatment. The mean of arch reintervention time was 8.7 ± 7.3 months (range, 2–27 months), and the angioplasty evaluation of the mean gradient 38.5 ± 15.5 mmHg dropped to the mean of 6.1 ± 4.5 mmHg gradient by balloon angioplasty. One case required stent implantation (9×17 mm^2 stent; Visi-pro, Medtronic) to the descending aorta after surgical reintervention. There were no cases of bronchial compression or aneurysm of the aortic arch. Patients requiring reintervention were presented in Table 5.

Of the remaining 15 patients (65.2%) in Group 1, two were lost to follow-up. Two patients underwent pulmonary debanding with VSD closure. Two underwent transcatheter pulmonary debanding, and remaining small muscular VSDs were followed up. One patient underwent the Yasui procedure, one underwent Glenn with Damus–Kaye–Stansel anastomosis, and one underwent pulmonary debanding with AVSD repair. All patients survived after the second stage operation and were in good clinical condition. The remaining six patients were under follow-up for the second stage operation.

5 | DISCUSSION

The surgical management of AAH with associated intracardiac anomalies is a challenge in neonates and infants. According to intracardiac anomalies, palliation or total repair in addition to arch repair is controversial. Moreover, significant proximal transverse aortic arch obstruction is rarer than the distal arch and isthmus hypoplasia, but almost always requires a surgical approach via a sternotomy.[1] In our cohort, all patients had tubular arch hypoplasia, so we performed all arch reconstructions via sternotomy and enlarged the aortic patches to the midportion of the ascending aorta. We attempted to perform total intracardiac repair in all biventricle patients, but in single ventricle patients and biventricle patients with preoperative comorbidities we performed pulmonary artery banding. From this point of view, BSA values during surgery were significantly lower in the palliation group and the number of patients weighing <2.5 kg was higher.

Several techniques and patch types have been proposed through the years for repair of aortic arch obstructions, and recurrent obstruction in the correspondence of the aortic arch surgery is the most frequently reported complication, with a hugely variable incidence from 2% to 38%.[5–7] Morales et al.[8] presented excellent results with 100% freedom from arch reintervention during a 5-year follow-up in neonates with interrupted aortic arch using the aortic arch advancement technique. On the other hand, Gaynor et al.[9] found no difference in the incidence of reintervention by patch reconstruction of the arch (20%) or resection with end-to-end anastomosis (25%). In our technique, the distal descending aorta was anastomosed to the

TABLE 3 Postoperative early outcomes

Outcome	Total ($n = 60$)	Palliation ($n = 23$)	Total correction ($n = 37$)	p value
Mechanical ventilation time, days, median (range)	5 (1–225)	5.5 (1–225)	5 (1–220)	0.56
ICU length of stay days, median (range)	11 (2–228)	15 (2–150)	10 (2–140)	0.45
Hospital length of stay, days, median (range)	19 (2–230)	19 (2–225)	19 (2–230)	0.93
Delayed sternal closure, *n* (%)	35 (58.3%)	12 (52.1%)	23 (62.1%)	0.44
Delayed sternal closure time, mean, days	4.3 ± 5.9	6.8 ± 8.7	2.7 ± 2.1	**0.044**
VAC theraphy, *n* (%)	5 (8.3%)	4 (17.3%)	1 (2.7%)	**0.045**
Bleeding reexploration, *n* (%)	3 (5%)	2 (8.7%)	1 (2.7%)	0.3
Sudden cardiac arrest, *n* (%)	4 (6.6%)	4 (17.3%)	0 (0%)	**0.009**
ECMO, *n* (%)	2 (3.3%)	0 (0%)	2 (5.4%)	0.25
Low cardiac output, *n* (%)	8 (13.3%)	6 (26%)	2 (5.4%)	**0.02**
Recoarctation, *n* (%)	13 (21.6%)	4 (17.3%)	9 (24.3%)	0.52
Chylothorax, *n* (%)	1 (1.6%)	0 (0%)	1 (2.7%)	0.42
Left diaphragm paralysis, *n* (%)	2 (3.3%)	0 (0%)	2 (5.4%)	0.25
Renal insufficiency, *n* (%)	3 (5%)	2 (8.7%)	1 (2.7%)	0.3
Prolonged (>7 days) mechanical ventilation, *n* (%)	23 (39.6%)	10 (45.4%)	13 (36.1%)	0.48
Tracheostomy, *n* (%)	4 (6.6%)	2 (8.7%)	2 (5.4%)	0.61
Prolonged (>15 days) ICU stay, *n* (%)	24 (40%)	12 (52.1%)	12 (32.4%)	0.12
Minor neurological events, *n* (%)	3 (5%)	1 (4.3%)	2 (5.4%)	0.85
Pericardial drain, *n* (%)	1 (1.6%)	0 (0%)	1 (2.7%)	0.42
In hospital mortality, *n* (%)	12 (20%)	8 (34.7%)	4 (10.8%)	**0.024**

Note: Bold values are statistically significant.
Abbreviations: ECMO, extracorporeal membrane oxygenation; VAC, vacuum-assisted closure.

aortic arch to form a native posterior wall, and patch augmentation was extended to the midportion of the ascending aorta. In our opinion, it is safer not to use too long and wide patches and to extend the patch. We preferred glutaraldehyde-treated autologous pericardium in 21 patients, bovine pericardium in 19 patients, and porcine pericardium in 20 patients as a patch. Rigid patches, in particular grafts, can cause stenosis by pushing the proximal and distal aorta.

All of our patients had tubular arch hypoplasia in this study, which was calculated to be a risk factor for recoarctation by Dodge-Khatami et al.[10] in a 40-year review of patients undergoing coarctation repair. Similarly, Bernabei et al.[7] presented a higher incidence of restenosis rate with neonatal arch hypoplasia compared with arch interruption or the hypoplastic left heart syndrome. Interestingly, all patients in that study underwent arch reconstruction with autologous pericardium. One- and three-year freedom from recoarctation was 93% and 69%, respectively, and the authors concluded that the use of autologous pericardium in aortic arch reconstruction procedures is valid and associated with an acceptable incidence of recurrent arch obstruction.[7] In our study, 10 (20.8%) patients required reintervention due to distal recoarctation, seven of them treated with successful aortic balloon angioplasty, but three required surgical reintervention. We performed resurgery via left thoracotomy in three patients using bovine or porcine pericardium. We observed only one (2%) proximal restenosis, probably due to the particular shape of the aortic arch, and we performed reoperation via median sternotomy. We believe that our low proximal restenosis rate depends on our technique that extends the patch to the middle of the ascending aorta. Furthermore, with the use of patch enlargement, we did not observe bronchi or trachea compression described as a possible complication of end-to-end anastomosis.[11]

The arch hypoplasia, interruption, or coarctation is efficiently dealt with at the time of the ASO while working through a median sternotomy. It is generally possible to perform a direct anastomosis for interruption or coarctation with mild tension on the anastomosis. On the other hand, the AAH requires an additional longitudinal patch-plasty. This has the advantage of enlarging the size of the ascending aorta, thus facilitating a tension-free neoaortic anastomosis

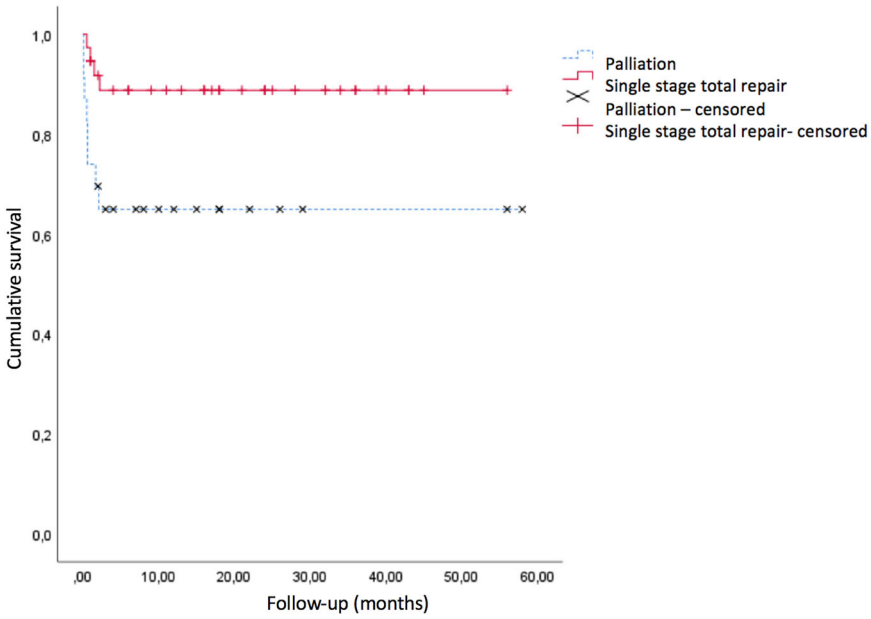

FIGURE 2 Kaplan–Meier survival for both groups undergoing aortic arch repair with 95% confidence interval at follow up

and achieves an aortic arch of adequate size. Huber et al. conducted a study on 22 patients who underwent ASO and arch repair with 18.1% overall mortality, and three patients required ECMO. Sixteen of 22 patients had arch hypoplasia, and the remaining patients had coarctation or IAA; Huber et al.[12] reported a 44% rate of arch reintervention. Vouhe's group described a 76% actuarial survival rate in 38 patients who underwent ASO with arch repair, mentioning arch reintervention in three patients (7.8%).[13] By contrast, Planche's group conducted a study on 67 patients who underwent one-stage repair with either end-to-end (35 patients) or patch enlargement of (32 patients) for AAO relief. Overall, actuarial survival in this series was 94% in the 32 patients and 75% in the 35 patients over 10 years, and reintervention was required in 15 patients (22%) for recoarctation.[14] In our cohort, 11 patients underwent ASO and arch repair with 18.8% overall mortality; one of them required ECMO, two patients died due to sepsis on the postoperative course, and only one patient (11%) required reintervention for recoarctation during the follow-up.

TABLE 4 Univariate analysis of risk factors for death

Risk factor	*p* value	HR (95% CI)
Palliation (PAB + arcus repair)	.019	0.44 (0.09–2)
Female	.98	-
Prematurity	<.001	4.67 (1.34–16.18)
Genetic syndrome or poor preoperative conditio	.234	-
Weight ≤2.5 kg	.116	-
Preoperative mechanical ventilatory support	.048	1.83 (0.52–6.39)
Functional single ventricle	.006	0.43 (0.1–1.86)

Abbreviations: CI, confidence interval; HR, hazard ratio; PAB, pulmonary artery banding.

Our mortality and reintervention rates were similar to the literature for this complicated group. We considered that the one-stage-repair of transposition complex and AAH showed good survival results for these highly complex congenital anomalies in high-volume hospitals.

More complex pathologies or functional single ventricle with arch hypoplasia showed high morbidity or mortality rates.[15] Historic outcomes of PAB + COA repair have been unfavorable and associated with high hospital death, reduced progression toward subsequent palliative stages, and overall low survival.[16,17] Franklin et al. found that survival after PAB + COA repair was 44% at 1 year and 22% at 5 years, and in another study, they found that Fontan candidacy was only 8%.[16,17] In our study, in Group 1 (PAB + arch repair), the mortality rate was 34.8% (8/27), and 62.5% (5/8) of them had functional single ventricle. On the other hand, Poirier et al. conducted a study on 37 children who had arch hypoplasia and biventricular hearts, and they reported no postoperative deaths with isolated AAH patients. However, the operative mortality for their total cohort of patients was 13.5%.[18] Karl and associates have reported similar results for the combined arch and intracardiac repair with a 13% operative mortality rate in a group of 15 infants who underwent a single-stage repair.[19] These mortality rates were similar to our single-stage repair group, with a mortality rate of 10.8%. In our study, only six cases with biventricular heart underwent isolated arch reconstruction with no operative mortality. Although the duration of CPB time and cross clamp time were significantly higher in the single-stage repair group, postoperative mortality, delayed sternal closure time, VAC treatment usage rate, sudden cardiac arrest rate, and postoperative low cardiac output rates were significantly higher in the palliation group. In our perspective, these results show the importance of single stage total repair, and the adverse effect of the palliation to the outcomes.

In neonates and some infants, low cardiac output can persist after coarctation repair due to preoperative LV dysfunction.[20] Lim

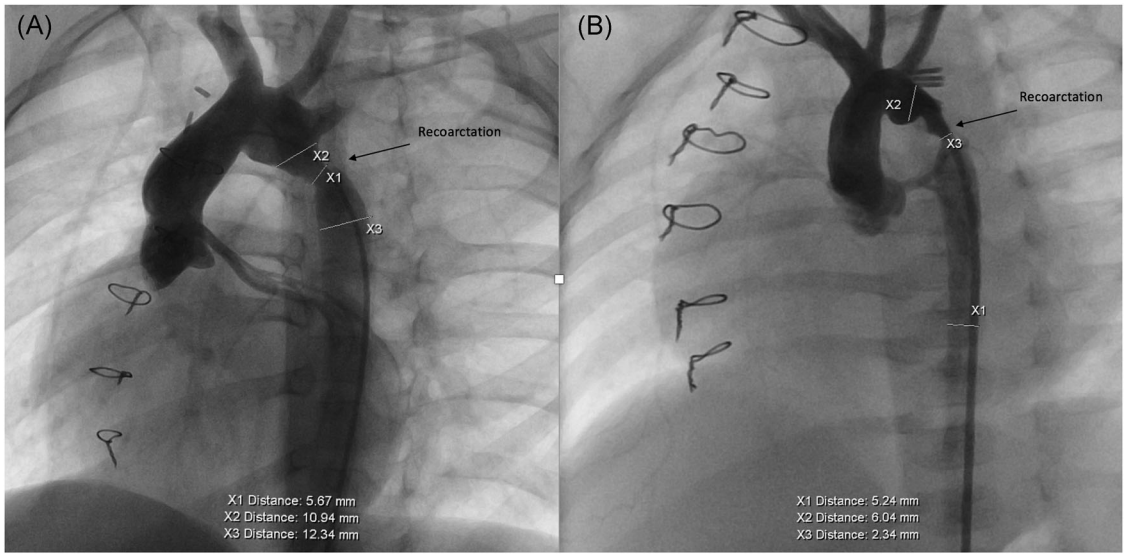

FIGURE 3 (A, B) Angiographic images of two patients with postoperative recoarctation

et al. conducted a study of 69 neonates or infants who underwent single-stage total repair of the aortic arch anomaly using regional perfusion. They reported that only four patients (5.7%) had postoperative low cardiac output syndrome, and all of them had poor preoperative conditions.[20] In our study, eight patients (13.3%) had postoperative low cardiac output syndrome, and this was significantly higher in our palliation group ($p = .022$). Among eight patients, seven required mechanical ventilatory support, and three of them had poor ventricular function requiring inotropic in the preoperative period. Only one patient did not need any support preoperatively, but he was also premature.

We only used a direct cannulation method through the innominate artery, even in the small neonates, instead of using a polytetrafluoroethylene graft. We considered it a basic and applicable method that did not conceal the surgical area. Moreover, there was no cannulation-related complication, such as stenosis or intimal injury in our cohort. Usually, the size of the innominate artery was adequate for cannulation, and we easily advanced the 8 Fr arterial cannula. We used ACP perfusion during the repair of AAH with a flow rate of 40–50 ml/kg/min to maintain a perfusion pressure of 50–60 mmHg, and all patients were monitored by NIRS. In our series, early postoperative minor neurologic complications as seizures occurred in three patients (5%), which is better than the reported rates (4–25%) in total circulatory arrest groups.[21] Any abnormality was not detected in these patients on computed tomography and electroencephalogram, and fortunately, all patients recovered entirely by antiepileptic therapy, but the long-term complications such as neurodevelopmental abnormalities need to be evaluated further.

The retrospective and single-center study design aspects are the main limitation of this study. Studies with multiple centers, including

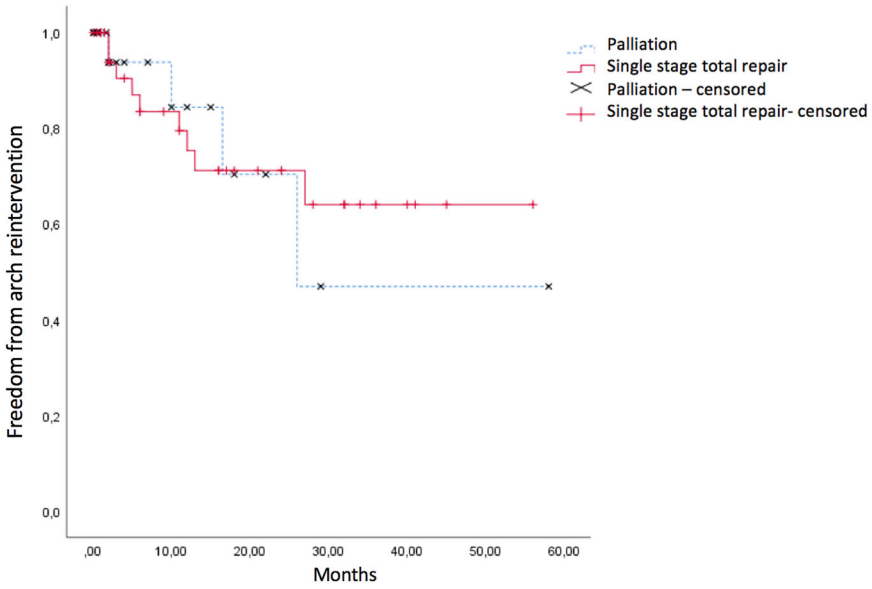

FIGURE 4 Kaplan–Meier curve showing the freedom from arch reintervention for both groups

TABLE 5 Patients requiring catheter or surgical reinterventions

Group	Age (days)	Weight (kg)	Preoperative proximal transerve arch *Z* score	Preoperative distal transerve arch *Z* score	Preoperative isthmic arch *Z* score	Comorbidities	Recoarctation localization	Associated procedure	Time to intervention (months)	Reintervention type
Palliation	120	5	−4	−1.9	1.3	-	Descending aorta (distal suture line)	PA banding + aortic valve comissurotomy	16.5	Balloon angioplasty
Total correction	35	3	−7	−3.7	−2.6	-	Descending aorta (distal suture line)	VSD closure	2	Surgery followed by stent implantation 1 year later
Total correction	28	3.4	−5.6	−2.9	−3.8	-	Descending aorta (distal suture line)	Arterial switch + VSD closure	3	Surgery
Palliation	12	2.8	−2.2	−0.4	−0.7	-	Descending aorta (distal suture line)	PA banding	10	Balloon angioplasty
Total correction	60	4.4	−2.4	−0.5	−1.1	-	Descending aorta (distal suture line)	-	12	Balloon angioplasty
Total correction	4	2.7	−5.3	−3.6	−5.9	Premature	Descending aorta (distal suture line)	-	6	Balloon angioplasty
Palliation	10	2.5	−7	−5.2	−5.8	-	Descending aorta (distal suture line)	PA banding	2	Surgery
Total correction	9	3	−7.4	−2.7	−6	Williams syndrome	Ascending aorta (proximal suture line)	VSD closure	27	Surgery
Total correction	28	3.5	−5.4	−3.4	−3.8	-	Descending aorta (distal suture line)	VSD closure	5	Balloon angioplasty
Total correction	31	3.5	−5.6	−3.4	−4	-	Descending aorta (distal suture line)	VSD closure	2	Balloon angioplasty
Total correction	41	3.6	−3.6	−1.7	−2.3	History of esophageal atresia surgery	Descending aorta (distal suture line)	VSD closure	11	Balloon angioplasty

Abbreviations: PA, pulmonary artery; VSD, ventricular septal defect.

various types of congenital heart disease and long-term outcomes, are needed to demonstrate the surgical results of AAH.

In conclusion, aortic arch reconstruction for tubular arch hypoplasia in neonates and infants with or without associated intracardiac lesions can be repaired by single-stage repair with acceptable early and midterm results. On the other hand, palliation with arch repair should be considered in biventricular neonates with significant preoperative comorbid conditions, and in single ventricle neonates.

CONFLICT OF INTERESTS

The authors declare that there are no conflict of interests.

ETHICS STATEMENT

The authors assert that all procedures contributing to this study comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008, and have been approved by the institutional committees (ATADEK ethics committee approval N°: 2020-2/6). The authors assert that all procedures contributing to this study comply with the ethical standards of the relevant national guides of care and have been approved by the institutional committee (ATADEK ethics committee approval N°: 2020-2/6).

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author, [author initials], upon reasonable request.

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