




Outcomes of surgery for coarctation of the aorta based on a new classification system

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Original Article

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Abstract

Objectives: To evaluate early- and long-term outcomes of the surgical treatment for coarctation of the aorta based on a new classification system. **Methods:** A retrospective clinical review of 111 patients with coarctation of the aorta who underwent surgery (March 2011 to August 2020) was performed. We categorised coarctation of the aorta into type I, with all three head vessels tightly packed; type II, with the left subclavian artery separated from the two other head vessels; and type III, with all three head vessels separated from one another. Each type included subtype a, with a short isthmic portion, and subtype b, with a long isthmic portion. **Results:** The median patient age and weight at operation were 8 (range, 1–1490) days and 3.2 (range, 1.9–18.5) kg, respectively. Extended end-to-end anastomosis was performed via sternotomy in 54, via thoracotomy in 12, end-to-side anastomosis in 31, autologous main pulmonary artery patch augmentation in 12, and modified end-to-end anastomosis combined with subclavian artery flap aortoplasty in two patients. There was one (0.9%) case of early mortality and 12 (10.8%) cases of post-operative complications. Two (1.8%) late deaths occurred during follow-up. Five (4.5%) patients underwent balloon dilatation and three (2.7%) underwent reoperation for restenosis of coarctation of the aorta. All patients with type Ia (21 patients, 18.9%) underwent extended end-to-end anastomosis via sternotomy or thoracotomy. **Conclusions:** According to the early and late outcomes observed in this study, surgical treatment of coarctation of the aorta using the new classification system could be safe and low risk.

The optimal surgical treatment for coarctation of the aorta remains controversial, in particular, in patients with tubular hypoplasia of the aortic arch. Therefore, diverse surgical techniques, including end-to-end anastomosis, subclavian artery flap aortoplasty, extended end-to-end anastomosis,^{1,2} end-to-side anastomosis,^{3,4} patch^{5,6} or conduit repair, and other alternative options,⁷ with or without cardiopulmonary bypass, have been used. We believe that the management of coarctation of the aorta should be based on the anatomical and morphological features of the aortic arch and isthmic portion. Therefore, we devised a surgical classification for coarctation of the aorta based on the anatomy of the aortic arch and morphology of the isthmic portion, as described in several studies,^{8–10} and managed coarctation of the aorta according to this classification. The aim of this study was to evaluate early- and long-term outcomes associated with the surgical treatment for coarctation of the aorta, chosen according to surgical strategies based on the coarctation of the aorta classification.

Patients and methods

Ethical statement

The Institutional Review Board of Pusan National University Yangsan Hospital approved the present study (IRB No. 05-2021-002, January 12, 2021), and the requirement for patient consent was waived owing to the retrospective data analysis nature of the study.

Patients and surgical classification of coarctation of the aorta

A retrospective clinical review of patients with coarctation of the aorta who underwent surgery from March 2011 to August 2020 was conducted. We excluded patients that presented with Taussig–Bing anomaly, transposition of the great arteries, truncus arteriosus, complex coarctation of the aorta requiring extra-anatomic repair, and double-lumen aortic arch.¹¹ Several classifications of coarctation of the aorta with a hypoplastic aortic arch have been previously published.^{8–10} This evidence suggests that coarctation of the aorta repair should be based primarily on the anatomical variations observed in the aortic arch and the morphology of

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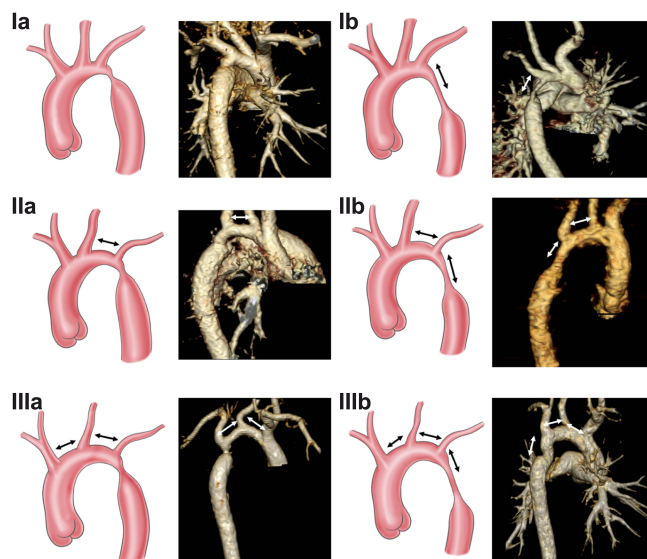


Figure 1. Each type of morphology of coarctation of the aorta as shown by preoperative computed tomography angiography.

the isthmic portion. Therefore, we devised a surgical classification for coarctation of the aorta that allows surgeons to choose a suitable surgical technique based on these anatomical characteristics.

Based on the anatomical features of the aortic arch, we proposed three types of coarctation of the aorta: type I in which all three head vessels are tightly packed; type II in which the left subclavian artery is separated from the other two head vessels; and type III in which all three head vessels are separated from one another. Each of these three types has subtypes according to the morphology of the isthmic portion: subtype (a) has a short isthmic portion and subtype (b) has a long isthmic portion. The aortic arch is divided into three segments. The proximal portion is between the innominate artery and left carotid artery, the distal portion is between the left carotid artery and left subclavian artery, and the isthmic portion is between the left subclavian artery and patent ductus arteriosus. The anatomic criteria for arch hypoplasia are a proximal portion of the aortic arch of $<60\%$ of the ascending aortic diameter and a distal portion of the aortic arch of $<50\%$ of the ascending aortic diameter^{12,13} or when the transverse aortic arch diameter is less than the standard as per the patient's weight in kilograms plus one (i.e., 5 mm for a 4 kg patient).¹⁴ Finally, six different types of coarctation of the aorta were assigned. Figure 1 shows each type of coarctation of the aorta morphology visualised using pre-operative CT angiography.

Surgical approach and techniques

All patients underwent single-stage repair through thoracotomy without a cardiopulmonary bypass or median sternotomy with a cardiopulmonary bypass. When we performed the procedure with a cardiopulmonary bypass for intracardiac anomaly or extensive aortic arch reconstruction using end-to-side anastomosis or main pulmonary artery patch augmentation, arterial cannulation was performed by grafting with a 3.5-mm polytetrafluoroethylene tube graft sutured to the innominate artery or our specially designed cannula tip since January 2015, as previously described.¹⁵ Another arterial cannula was inserted into the descending aorta through the patent ductus arteriosus, if necessary. Intracardiac repair was performed with standard bicaval cannulation with cold

blood cardioplegia, and antegrade selective cerebral perfusion through the innominate artery was conducted during arch reconstruction while monitoring the cerebral oxygenation.

Procedure type for each patient with coarctation of the aorta was selected according to our classification system. When the aortic arch showed that all three head vessels were tightly packed (type I), surgery was performed using extended end-to-end anastomosis via thoracotomy or sternotomy. If the patient showed a long isthmic portion (subtype b), end-to-side anastomosis could be considered. When the aortic arch showed that the left subclavian artery was separated from the other two head vessels (type II) or when all three head vessels were separated from one another (type III), surgery was performed according to the size of the transverse aortic arch segment. Namely, when the proximal aortic arch was hypoplastic, main pulmonary artery patch augmentation was performed; when the distal aortic arch was hypoplastic, end-to-side anastomosis was performed; and when the proximal and distal aortic arches were sizable, the patient underwent extended end-to-end anastomosis or end-to-side anastomosis depending on the subtype. Therefore, extended end-to-end anastomosis was performed for subtype a, and either extended end-to-end anastomosis or end-to-side anastomosis was performed for subtype b (Supplementary Figure 1). When we performed the main pulmonary artery patch augmentation, a patch of the main pulmonary artery was harvested from the anterior wall of the main pulmonary artery, from the valve commissures to the pulmonary bifurcation while paying particular attention to ensure that we stayed a distance away from both pulmonary artery branches' ostia. Afterward, an incision was made along the inferior aspect of the aortic arch, usually between the innominate artery and descending aorta, and when posterior anastomosis was completed, the remaining anterior aspect of the aortic arch was supplemented with the autologous main pulmonary artery patch harvested.^{6,16,17}

A surgical alternative was available for patients who had a long isthmic portion (subtype b) and a sizable transverse aortic arch regardless of the anatomical features of the aortic arch. Recently, we performed modified end-to-end anastomosis combined with subclavian artery flap aortoplasty in two patients with pre-operative airway compression, as previously described.^{7,18} The operation was performed through a left thoracotomy in the fourth intercostal space, without cardiopulmonary bypass. After extensive mobilisation of the aortic arch and descending aorta, subclavian artery flap aortoplasty was performed under descending aortic perfusion through the patent ductus arteriosus, and afterward, end-to-end anastomosis was performed. Because the subclavian artery flap was usually longer than the isthmic portion, some parts of the subclavian artery flap could be used to augment the posterior part of the bevelled proximal descending aorta (Fig 2).

Early mortality was defined as death during hospitalisation or within 30 days after surgery. Late mortality was defined as death occurring >30 days postoperatively and after discharge from the hospital. Aortic arch reintervention was defined as balloon dilation or reoperation of the aortic arch.⁵

Statistical analyses

Data were collected and managed using Microsoft Excel 2021 and R software version 3.5.3 (R Foundation for Statistical Computing). Descriptive statistics were derived for the study population. Continuous variables are presented as median with range or mean \pm standard deviation, depending on the distribution patterns, which were tested using the Kolmogorov–Smirnov test.

Table 1. Concomitant procedures

Procedure	Number of patients	%
VSD closure	68	61.3
PA banding	4	3.6
AV valvotomy	3	2.7
LV diverticulectomy	1	0.9
PV repair	1	0.9
Posterior aortopexy	1	0.9
Supra-mitral ring resection	1	0.9
Total	79	

AV = aortic valve; LV = left ventricle; PA = pulmonary artery; PV = pulmonary valve; VSD = ventricular septal defect.

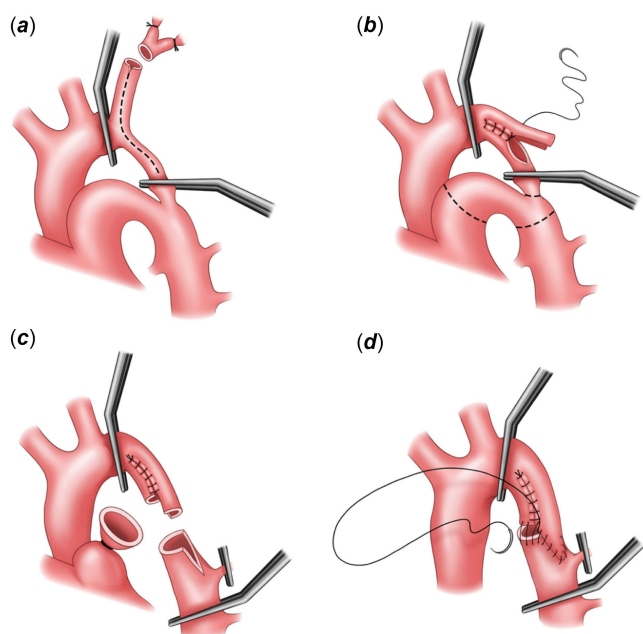


Figure 2. Modified end-to-end anastomosis (EEA) combined with subclavian artery (SCA) flap aortoplasty. (a and b) SCA flap aortoplasty was performed under descending aortic perfusion through the patent ductus arteriosus. (c and d) EEA was performed. Part of the SCA flap was used to augment the posterior part of the bevelled proximal descending aorta.

Categorical variables are presented as counts and percentages. Freedom from reintervention was determined using the Kaplan-Meier curve, and a 95% confidence interval was applied.

Results

Overall, 111 consecutive patients (73 male individuals, 65.8%) were included in the study. The median age and body weight at the time of surgery were 8 (range, 1–1490, IQR: 13) days and 3.21 (range, 1.9–18.5, IQR: 0.8) kg, respectively. The anatomical type of each patient with coarctation of the aorta is described in Supplementary Table 1. The most common types of coarctation of the aorta were type IIa (37 patients, 33%), type Ib (28 patients, 25.2%), and type Ia (21 patients, 18.9%). A total of 79 concomitant procedures were performed in 76 (68.5%) patients, and ventricular septal defect closure was the most frequently performed procedure (68 patients, 61.3%) (Table 1). Table 2 presents the operative

techniques used for patients with each anatomical type. All except two patients (109 patients, 98.2%) underwent coarctation of the aorta repair following our surgical strategy. The surgical strategy was not followed for the two remaining patients (marked with “*” in Table 2). Instead, the two patients underwent main pulmonary artery patch augmentation because of their relatively old age (4 years and 1 month and 19 months, respectively), which made it difficult to mobilise the aortic arch and descending aorta extensively, precluding tension-free anastomosis without patch augmentation. For the main pulmonary artery patch augmentation technique, the defect at the main pulmonary artery was usually repaired with glutaraldehyde-treated autologous pericardium patch except in two patients (one, using the remaining autologous main pulmonary artery patch because of the very large size of the main pulmonary artery patch, and the other one, bovine pericardium patch for the redo-operation).

Early results

There was one surgery-related mortality (0.9%). The patient showed pre-operative left ventricular dysfunction and died of post-operative low cardiac output and sepsis. Post-operative complications occurred in 12 patients (10.8%). These included diaphragmatic palsy (n = 7), vocal cord palsy (n = 6), chylothorax (n = 5), reintubation (n = 4), atrioventricular block (n = 4), left main bronchus obstruction (n = 2), and prolonged ventilator care (n = 1).

Late results

The mean follow-up duration was 43.9 ± 29.8 months. There were two (1.8%) late deaths, although none were related to aortic arch problems. All causes of death were unknown (two patients died suddenly at home). One patient had epilepsy and cerebral palsy, while the other was a syndromic patient.

During the follow-up period, five (4.5%) patients required balloon dilation and three (2.7%) required surgical reoperation of the aortic arch. Overall, the rates of freedom from reintervention (surgery or balloon dilation) were 98.1%, 93.9%, and 92.5% at 6 months, 1 year, and 5 years after surgery, respectively (Supplementary Figure 2).

Modified end-to-end anastomosis combined with subclavian artery flap aortoplasty

Two patients underwent modified end-to-end anastomosis combined with subclavian artery flap aortoplasty. The first patient was a premature baby born at 33 weeks and 3 days of gestation. The aortic arch had a type IIb morphology (the left subclavian artery was separated from the other two head vessels with a long isthmic portion) and a sizable transverse aortic arch. In addition, the patient showed compression of the tracheal bronchus and the left main bronchus. Therefore, we performed modified end-to-end anastomosis combined with subclavian artery flap aortoplasty at 55 days of age (4.15 kg) and obtained good surgical results. The second patient was also a premature baby born at 32 weeks and 1 day of gestational age. The patient had a type Ib aortic arch (all three head vessels were tightly packed with a long isthmic portion) and a sizable transverse aortic arch. Pre-operative airway compression and an anteriorly positioned descending aorta were identified. We decided to perform modified end-to-end anastomosis combined with subclavian artery flap aortoplasty at

Table 2. Surgical techniques for patients with each anatomical type according to our new classification

Anatomical type	EEEE via sternotomy	EEEE via thoracotomy	ESA	MPA patch augmentation	Modified EEA combined with SCA flap
Ia	16	5	0	0	0
Ib	20	2	3	2*	1
IIa	12	2	19	4	0
IIb	5	2	3	4	1
IIIa	1	0	5	2	0
IIIb	1	1	0	0	0
Total	55 (49.5%)	12 (10.8%)	30 (27%)	12 (10.8%)	2 (1.8%)

EEA = end-to-end anastomosis; EEEA = extended end-to-end anastomosis; ESA = end-to-side anastomosis; MPA = main pulmonary artery; SCA = subclavian artery.

*Indicates patients who did not follow our surgical strategy.

28 days of age (2.15 kg), and we obtained satisfactory results (Supplementary Figure 3).

Discussion

In this study, we reviewed patients who had undergone surgery for coarctation of the aorta, and whose defects were classified coarctation of the aorta based on the anatomy and morphology of the aortic arch and isthmic portion, respectively. Our findings show that a surgical approach to coarctation of the aorta based on our new classification system could be a safe and low-risk option in terms of short- and long-term outcomes. In addition, the proposed surgical classification for coarctation of the aorta may allow surgeons to choose an appropriate surgical approach based on the patient's anatomical characteristics.

Since the first coarctation of the aorta repair by Crafoord and Gross,^{19,20} controversies have remained regarding optimal surgical techniques, in particular, for patients with tubular hypoplasia of the aortic arch. In 1983, Waldman et al.²¹ stated that the choice of a surgical technique for coarctation of the aorta should be based on the patient's anatomical features rather than the surgeon's preference. Similarly, we believe that the management of coarctation of the aorta should be based on the anatomical features of the aortic arch and the morphology of the isthmic portion. Although the anatomical characteristics of coarctation of the aorta are diverse, we identified some typical forms of the aortic arch and isthmic portion, and proposed a classification system based on these patterns, which overlap with previously identified patterns, aiming to support surgeons in selecting the optimum operative approach.^{8–10}

Surgical approach according to the classification for coarctation of the aorta

Decision-making for patients with coarctation of the aorta was performed according to our classification system. The process began with an evaluation of the anatomical characteristics of coarctation of the aorta, which included the anatomical features of the aortic arch and morphology of the isthmic portion. Different surgical approaches are recommended in different situations. First, when a patient presents with tightly packed head vessels with a short isthmic portion (type Ia), the length of the aortic arch is relatively short, and extended end-to-end anastomosis could be performed without excessive tension at the anastomosis site. However, a patient with subtype b (a long isthmic portion) could have a long aortic arch, including the isthmic portion, potentially making end-to-side anastomosis a suitable alternative. Second,

when the left subclavian artery is separated from the other two head vessels (type II) or when all three head vessels are separated from one another (type III), surgery should be performed according to the size of the transverse aortic arch segment. Patients with type II or type III aortic arch morphology have a relatively long aortic arch. Therefore, when the proximal aortic arch is hypoplastic, an enlargement of the proximal aortic arch can be effectively achieved using main pulmonary artery patch augmentation. When end-to-side anastomosis are performed, we usually incise the lesser curvature of the aortic arch up to the site of the innominate artery; in contrast, some surgeons have performed end-to-side anastomosis between the distal ascending aorta and extensively mobilised the descending aorta, which is similar to the method used in interrupted aortic arch repair.^{4,14,22} However, this aggressive end-to-side anastomosis method between the ascending and descending aorta could have some drawbacks, including an acute-angled aortic arch morphology such as a Gothic arch, excessive tension on the anastomosis site even after full mobilisation, and possible left main bronchus compression after surgery.^{4,22} Overall, this evidence suggests that the main pulmonary artery patch augmentation should be performed when a proximal aortic arch enlargement is required. End-to-side anastomosis can be performed when the distal aortic arch is hypoplastic. Finally, when the proximal and distal aortic arches are sizable, extended end-to-end anastomosis or end-to-side anastomosis can be performed depending on the morphology of the isthmic portion, as in type I coarctation of the aorta cases.

End-to-end anastomosis combined with subclavian artery flap aortoplasty as a surgical alternative

When the patient shows a long isthmic portion (subtype b) and a sizable transverse aortic arch, regardless of the aortic arch type, end-to-end anastomosis combined with subclavian artery flap aortoplasty can be performed as a surgical alternative. Amato et al.⁸ used a subclavian artery flap turnaround with end-to-end anastomosis for patients with coarctation of the aorta and isthmic hypoplasia. Hovaguimian et al.¹⁸ reported good results from modified end-to-end anastomosis with subclavian artery flap angioplasty, and Suzuki et al.⁷ introduced modified end-to-end anastomosis combined with subclavian artery flap aortoplasty for the repair of coarctation of the aorta with extended hypoplasia of the aortic isthmus. Our modified end-to-end anastomosis combined with subclavian artery flap aortoplasty has the following advantages. (1) Our method can save almost the entire isthmus and preserve the lesser curvature of the aortic arch, thereby preserving the natural geometry and length of the aortic arch after surgery.

(2) Our modification of the combined technique requires a short period of blood flow interruption through the descending aorta using descending aortic perfusion via the patent ductus arteriosus. In our study, we applied this modified method in the treatment of two patients with pre-operative airway compression and achieved good results.

Study limitations

This study had some limitations. First, it was a single-centre retrospective analysis. Therefore, selection or unidentified confounding bias may have influenced the results. The indications for the surgical procedures for each anatomical type were determined by the surgeon in the operative field according to the patient's anatomical characteristics. Second, there was no proper comparative group in this study, and the follow-up duration was relatively short (mean, 43.9 ± 29.8 months), thus providing insufficient time to evaluate late hypertension or restenosis of coarctation of the aorta. A longer follow-up period and prospective comparative study are required to determine the optimal surgical treatment strategy for coarctation of the aorta in the future.

The surgical approach to coarctation of the aorta, selected based on our new classification system, could be safe and low-risk, leading to good short- and long-term outcomes. In patients with a long isthmic portion (subtype b) and sizable transverse aortic arch, modified end-to-end anastomosis combined with subclavian artery flap aortoplasty could be a good surgical alternative. Finally, when performing coarctation of the aorta surgery, the choice of a surgical technique should be based on the anatomy of the aortic arch instead of the surgeon's preference.²¹

Supplementary material. To view supplementary material for this article, please visit <https://doi.org/10.1017/S104795112300104X>

Data availability statement. The data underlying this article will be shared upon a reasonable request to the corresponding author.

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Si Chan Sung: validation, supervision, writing—reviewing, and editing

Hyoung Doo Lee: validation, supervision

Hoon Ko: investigation, resources

Joung-Hee Byun: investigation, resources

Taehong Kim: investigation, resources

Chanyoung Chung: investigation, resources, data curation, software

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Ethical standards. The authors assert that all procedures contributing to this work 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 has been approved by the institutional committees (Pusan National University Yangsan Hospital, IRB No. 05-2021-002, January 12, 2021).

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