Arcus Aorta Reconstructions in Neonates and Early Infants

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ABSTRACT

Introduction: Various instances of aortic arch hypoplasia (AAH) with a wide spectrum of severity have been observed in the neonatal patients. The purpose of this paper is to discuss the aortic arch reconstruction in the coarctation of the aorta and AAH while focusing on the surgical decisions and patient management for the neonatal and infant patients.

Patients and Methods: We performed a retrospective review of 15 patients who underwent aortic arch reconstruction. Importantly, we investigated postoperative outcomes with associated complex congenital heart diseases.

Results: The median age and mean weight of the patients were 74 days (range: 4-306 days) and 4192 ± 1253 gram, respectively. The surgical correction of aortic arch congenital abnormalities was performed under selective antegrade cerebral perfusion in all the 15 patients. All the reconstructions were performed with pericardial patch, and the coarcted segment of the aorta was completely resected in seven of the patients. One-stage repair was performed in the seven of the patients. The mean follow-up time was 13.7 ± 9.3 months. In one patient, transcatheter balloon angioplasty was performed for re-coarctation six months after operation. There was one early and one late mortality.

Conclusion: Aortic arch reconstruction can be performed with a low mortality and morbidity in the newborn and infants.

Key Words: Arch of the aorta; reconstructive surgical procedure; aortic arch reconstruction; neonate

Yenidoğan ve Bebeklik Döneminde Arkus Aorta Rekonstrüksiyonları

ÖZET

Giriş: Aortik ark hipoplazisinin çeşitliliği yenidoğan hastalarda geniş bir spektrumda görülebilir. Bu yazıda koarktasyon ve hipoplastik aortik ark tanılı hastalarda, aortik ark rekonstrüksiyonu tartışılacak, yenidoğan ve bebek hastalarda cerrahi kararlara ve hasta yönetimine odaklanılacaktır.

Hastalar ve Yöntem: Aortik ark rekonstrüksiyonu yapılan 15 hasta retrospektif olarak incelendi. Beraberinde komplike doğumsal kalp hastalıkları bulunan bu hastalarda operasyon sonrası cerrahi sonuçları araştırıldı.

Bulgular: Hastaların ortanca yaşı 74 gün (4-306 gün) ve ortalama ağırlığı 4192 ± 1253 gram idi. Aortik ark doğumsal hastalığının cerrahi tedavisi, 15 hastada antegrad serebral perfüzyon ile yapılmıştır. Bütün hastalarda aortik rekonstrüksiyon perikardiyal yama ile yapılmış ve yedi hastada koarkte segment tamamen rezeke edilmiştir. Hastaların yedisinde tek aşamalı onarım gerçekleştirilmiştir. Ortalama takip süresi 13.7 ± 9.3 aydı. Bir hastaya tekrar gelişen aort koarktasyonu nedeniyle ameliyat sonrası altıncı ayda balon anjiyoplasti uygulanmıştır. Takip sırasında bir erken ve bir geç mortalite saptanmıştır.

Sonuç: Yenidoğan ve bebeklerde aortik ark rekonstrüksiyonları düşük mortalite ve morbidite oranlarıyla yapılabilmektedir.

Anahtar Kelimeler: Arkus aorta hipoplazisi; cerrahi düzeltme

INTRODUCTION

Aortic arch hypoplasia (AAH) with or without coarctation of the aorta (CoA) can be defined as the developmental impairment of the arcus aorta in the embryological period and is a relatively common congenital anomaly with a heterogeneous clinical presentation[(1)]. Anatomic spectrum can vary from a mild isolated CoA to severe hypoplasia of the left heart structures[(2)]. Although a vast majority of cases have only shown an isolated ventricular septal defect (VSD), several more complex congenital intracardiac anomalies may include double

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outlet right ventricle, transposition of great arteries, atrioventricular septal defect, univentricular heart, and so on[1-3]. An early treatment may be needed because of the severity of hypoplasia and/or associated intracardiac defects. Several surgical techniques have been presented in the literature; however, patch aortoplasty and extended end-to-end anastomosis (EEEA) are the most commonly used techniques[1,3-5]. However, even with better surgical techniques for the neonates, additional structural anomalies or comorbidities such as the development of restenosis at the site of aortic anastomosis, left ventricular outflow tract obstruction, and left bronchial compression are still some risk factors for worse outcomes[4]. In this sight of view, every patient should be preoperatively and individually evaluated in detail for the severity of hypoplasia, its associated complex heart defects, and other risk factors because successful aortic arch reconstruction (AAR) is a critical component for an unobstructed systemic blood flow, especially in the setting of single ventricle physiology.

In this study, we retrospectively analyzed the outcomes of cases with the reconstructed aortic arch. The purpose of the study is to share our recent experiences about the surgical strategies and early outcomes of the patients with associated complex congenital heart diseases.

PATIENTS and METHODS

We retrospectively evaluated the data of the patients who were operated in our pediatric cardiovascular surgery clinic between November 2017 and April 2020. We included 15 neonates and early infants under the age of one year with the diagnosis of arcus aorta hypoplasia in association with different congenital heart defects in this study. These participants underwent AAR surgery with or without additional corrective or palliative heart surgery. Additionally, we excluded patients with CoA without arch hypoplasia, hypoplastic left heart syndrome, aortic arch interruption, and truncus arteriosus in this study. We recorded and analyzed the demographic data, operation notes, echocardiography, and radiological imaging reports of the patients. Importantly, we obtained the Institutional ethics committee's approval for our retrospective study (Date: June 9, 2020, Number: 905).

Cardiac and Vascular Evaluation

We performed a detailed evaluation of the patients in terms of preoperative and postoperative cardiac findings, physical examination, echocardiography, computed tomography (CT), and/or conventional catheter angiography.

Echocardiographic evaluations of the patients were performed with GE Vivid S6 (GE Healthcare Vingmed Ultrasound AS, Horten, Norway) 6S-RS sector probe (2.5-7.0 MHz) based on the "Guidelines and Standards for Performance of a Pediatric Echocardiogram" of the American Society of Echocardiography[6]. Four chamber views for left ventricle end diastolic dimension and mitral valve annulus; long axis views for aortic valve (AV) annulus and ascending aorta; suprasternal notch view for transvers aorta, distal arcus aorta, and isthmus were used for the evaluation and measurements of dimensions.

Three-dimension CT angiogram scans were obtained with Siemens Somatom Sensation Cardiac 64 device (Siemens Healthcare, Forchheim, Germany). Measurements of ascending aorta, transverse aorta (middle part of transverse arcus between truncus brachiocephalicus and left carotid artery), distal arcus aorta (immediately proximal to the part where the left carotid artery is separated), and isthmus [arch between the left subclavian and insertion of the patent ductus arteriosus (PDA)] were made in the axial and sagittal sections[7].

Conventional catheter angiography was performed to make an accurate diagnosis of the complex congenital heart defects. Additional CT scans were not performed to avoid additional radiation exposure in two patients. In conventional catheter angiography, a left view of 90º or left oblique angles of 45º were used to measure the required dimensions; data of two patients were obtained exclusively via this imaging technique.

Operative Technique

All operations were performed via standard median sternotomy. Arch vessels, both pulmonary arteries, the descending thoracic aorta, and a PDA (if present) were carefully mobilized. The aorta was cannulated with an 8Fr or 10Fr pediatric arterial cannula. In one patient, (ductal dependent) ductus arteriosus was cannulated with the 6Fr aortic canula for the lower body perfusion during the cooling phase. When concomitant intracardiac repair was necessary, venous cannulation was performed via the right atrium or with bicaval cannulation. Total circulatory arrest was achieved at 24°C along with the administration of antegrade cardioplegia. If necessary, interatrial septum was resected and selective antegrade cerebral perfusion (SACP) was initiated. The ductus arteriosus was completely resected in all the patients. The coarcted segment of the aorta was completely resected in seven of the patients. The minor curvature of the arcus aorta was incised through the mid portion of the ascending aorta. The posterior walls of the arcus and the descending aorta were sutured. The anterior wall of the aorta was reconstructed with pericardial patch in all the patients. During the rewarming period, concomitant intracardiac repairs were performed under the aortic cross-clamp (X-clamp). The aortic cross-clamp was released, and the patient was weaned from cardiopulmonary bypass (CPB). Modified ultrafiltration was performed in all the cases.

RESULTS

The median age and mean weight of the patients were 74 days (range: 4-306 days) and 4192 ± 1253 gr, respectively (Table 1). There were six neonates and nine infants in the patient population. Tables 2 and 3 show the Z-scores of cardiac and aortic structures along with associated complex heart defects. Aortic arch repair was performed by a curved porcine pericardium in eight of the patients, whereas it was performed with the autologous pericardium in six of the patients. Associated procedures were VSD closure in two neonates, arterial switch operation and VSD closure in two neonates, tunnel-type VSD closure and subaortic conus resection in one infant, infra-cardiac-type total anomalous pulmonary venous drainage and atrial septectomy in one neonate, pulmonary banding (PB) in eight patients (two of them were univentricular patients and two of them had multiple VSD), complete atrioventricular septal defect repair and subaortic stenosis resection were performed in one infant, and aortic commissurotomy was performed in one neonate. The mean follow-up time was 13.7 ± 9.3 months. There was one early mortality because of the low cardiac output at the borderline left ventricle (LV) in the patient and one late mortality in a patient with LV hypoplasia and VSD-dependent systemic circulation due to bulboventricular foramen restriction five months after the initial operation. Two patients required three reinterventions: one surgical and two catheter-based. Catheter-based reinterventions were applied on the same patient. First, catheter-based reintervention was applied because of aortic arch recoarctation, which was managed successfully by transcatheter balloon angioplasty six months after the operation. The second one was applied 18 months after the operation for the relief of the pulmonary banding site. Surgical reintervention, which was performed 24 days after the first operation, was a pulmonary rebanding due to inadequate PB.

DISCUSSION

AAH, which can be described as the congenital underdevelopment of aortic arch, may be associated with a wide spectrum of congenital heart diseases, and affect mortality and morbidity in the course of the disease or during the treatment based on

Table 1. Patient characteristics and operative data

Variable		Result
Age (days)	Median (min-max)	74 (4-306)
Weight (grams)	Mean ± SD	4192 ± 1253
Gender-male	n (%)	11 (73)
Extubation time (days)	Median (min-max)	4.5 (1-22)
ICU stay (days)	Median (min-max)	6 (2-51)
Hospital stay (days)	Mean ± SD	23 ± 13
Antegrade cerebral perfusion (minutes)	Mean ± SD	32 ± 10
CPB (minutes)	Mean ± SD	129 ± 43
X-clamp (minutes)	Mean ± SD	82 ± 44

SD: Standard deviation, ICU: Intensive care unit, CPB: Cardiopulmonary bypass, X-clamp: Aortic cross-clamp.

Table 2. Aortic arch and cardiac measurement Z-scores

Variables (Z-scores)	Mean ± SD (range)
AV annulus	-0.2 ±1.4 (-3 to 2)
Ascending aorta	-0.9 ± 1.6 (-3 to 0.8)
Transvers aortic arch	-3.9 ± 0.9 (-6 to -3)
Distal aortic arch	-2.8 ± 1.3 (-5.7 to 0.3)
Aortic isthmus	-3.4 ± 1 (-6.3 to -1.6)
LVEDd	0.14 ± 2.7 (-52 to 5.5)
MV annulus	-0.6 ± 1.9 (-5.6 to 2.1)

SD: Standard deviation, AV: Aortic valve, LVEDd: Left ventricle end diastolic dimension, MV: Mitral valve.

Table 3. Associated cardiac defects

Associated heart defects	n (%)
VSD (all)	9 (60)
TGA	3 (20)
LVOTO	2 (13)
cAVSD	3 (20)
DORV	1 (6)
TAPVD	1 (6)
DILV	1 (6)

VSD: Ventricular septal defect, TGA: Transposition of great arteries, LVOTO: Left ventricular outflow tract obstruction, cAVSD: Complete atrioventricular septal defect, DORV: Double outlet right ventricle, TAPVD: Total anomalous pulmonary venous drainage, DILV: Double inlet left ventricle.

the adequate systemic circulation. Several descriptions of AAH have been made before and were accepted in time; however, its diagnostic criteria may vary and is still considered controversial. This issue has an extreme importance because there is always a radical change in the decisions for using a particular surgical technique. As for AAH, surgical repair is performed using CPB via median sternotomy instead of thoracotomy. In 1992, Karl et al assumed the aortic arch to be hypoplastic if the transverse arch diameter was less than "the patient's weight in kilograms +1 mm"[8]. Backer and Mavroudis described isthmus hypoplasia as the condition wherein the isthmus is less than 40% of the diameter of the ascending aorta and when arch hypoplasia as proximal or distal transverse arch is less than 60% or 50% of the diameter of the ascending aorta, respectively, in "Congenital Heart Surgery Nomenclature and Database Project"[7]. Poncelet et al. used Z-scores and defined distal arch hypoplasia with a Z-score of less than -3 in their study[3]. An echocardiographic measurement formula "transvers arcus diameter divided to descending aorta diameter" was recommended by Rao et al, and a ratio of less than 0.65 was suggested to be significant[9]. Langley et al used a combination of three criteria, but they assumed that the Z-score of aortic arch diameter is less than -2.0 for hypoplasia[10]. In our study, we used Z-score of transvers aorta diameter to be less than -3.0 as an indication for reconstruction, and this Z-score ranged from -3 to -6. The patient with a Z-score of -6 also had hypoplastic left heart structures. With this patient population, all of our patients were treated with patch aortoplasty via median sternotomy. Besides, the surgical decision requires a careful assessment of the arch dimensions and anatomy. We usually used three-dimensional CT to evaluate the arcus. The measurements of two patients were obtained from conventional catheter angiography, which was performed for the evaluation of complex intracardiac pathology to avoid additional radiation.

Surgical techniques may affect early morbidities, mortality, as well as mid- and late-term reintervention rates. Resection and EEEA along with end-to-side anastomosis techniques are widely used in mild to moderate cases; however, median sternotomy and CPB are usually needed in severe hypoplasia[11]. However, reintervention rates are lower in sternotomy. Thulzer et al. reported no mortality with EEEA, but a reintervention rate of nearly 10% in ten years[11]. In a study from Zurich, Dave et al used both autologous pericardial augmentation patch for the roof of the intervening arch between the left carotid and left subclavian arteries, and EEEA via left posterior thoracotomy, but their reintervention requirement rate was relatively high (about 20%)[12]. All of our patients were operated with median sternotomy and CPB in consideration with the severity of arch hypoplasia. Only one patient experienced reintervention as transcatheter balloon angioplasty at the postoperative sixth month. The resection of coarctated segment was not performed in this patient's surgery, and need for reintervention was considered because of this technique. These results support the previous consideration that the resection of coarctation together with the direct anastomosis of the adjacent aortic ends and anterior aortic patch augmentation is probably an optimal technique for achieving a low incidence of re-coarctation[13].

Patch aortoplasty is a widely preferred surgery in the patients with AAH. Several materials such as synthetic materials, porcine or autologous pericardium, and pulmonary autograft have been used in this technique. Synthetic patches (Dacron, etc.) have shown technical difficulties in neonates and carry a risk of aortic aneurysm[14]. Biological materials such as autologous pericardium have relatively higher recurrence and reintervention rates. Bernabei et al. reported a 28.2% recurrence rate of the aortic arch obstruction postoperatively within months[15]. Reintervention rates of patients operated with an autologous pulmonary patch was reported to be low[1,4]. Contrarily, the use of autologous pericardium was considered to be a risk factor for re-coarctation with a rate of 26%[13]. In our study, arch repair was performed by a curved porcine pericardium in eight patients, whereas it was performed with the autologous peri-

cardium in seven of the patients. Reintervention rate was 6.2% with one patient, and it was lower than that was reported in the literature. Porcine pericardial patch was used in the surgery of this patient.

The protection of cerebral and visceral functions during aortic arch surgery is still a challenging issue. Traditional method, deep hypothermic circulatory arrest (DHCA), reduces the metabolic requisition of tissues including the central nervous system, but has shown some severe side effects such as multiorgan failure, coagulopathy, and increased inflammatory response[16]. Selective antegrade cerebral perfusion technique can be used to continue cerebral perfusion and reduce neurological complications. Kornilov et al. compared the two perfusion techniques, DHCA and SACP, and found neurological complication rates of 30.8% and 5.9%, respectively, thereby showing with SACP exhibited significantly better results[16]. We performed SACP at 24°C in all of our patients following the recent suggestions and did not experience any neurological complications in the early and midterm results[17].

Two patients from our study died later. One in early and the other one in midterm period, postoperative 16th day and 5th month, respectively. The first patient had borderline left heart structures with Z-scores of -3.5 for LV and -2.5 for MV. Management of the "borderline" left heart remains still a challenge. Plymale et al. suggested an echocardiographic assessment before surgery. They found that failed biventricular repair or reintervention is expected to be more common when the Z-scores of AV and MV were less than -2.5. They also reported the combination of MV and tricuspid valve ratio of less than 0.66 with an AV annulus Z-score of less than -3 can strongly predict biventricular circulation failure[18]. In our patient, AV Z-score was -3.3, the Z-scores of LV and MV were -3.4 and -2.5, respectively. After, impaired cardiac output, extra-corporeal membrane oxygenation (ECMO) support was provided to the patient, but cardiac hemodynamic findings did not improve and we were not able to do ECMO weaning. The patient died on the 16th day after ECMO initiation. The second patient died on the fifth postoperative month. The patient had very hypoplastic left heart structures with LV and MV Z-scores of -5 and -5.6, respectively, and a VSD providing systemic blood flow. As first stage, AAR and PB were performed, and the second stage was planned as cavo-pulmonary shunt at suitable age and weight. But unfortunately, the patient did not oriented routine postoperative visits and cardiac controls. He applied to the hospital at the age of five months with severe heart failure symptoms and a very bad clinical condition due to the restriction of bulboventricular foramen.

In conclusion, arcus aorta reconstructions can be performed with low mortality and morbidity in the very early stages of childhood. To achieve this goal, patients should be preoperatively evaluated in detail in terms of aortic measures and anatomy, associated complex heart defects, and especially competency of other left heart structures to achieve an adequate systemic circulation. Experience and practice will improve by further studies in this field.

Ethics Committee Approval: Ethics committee approval was received for this study from the Istanbul Yeni Yuzyil University Gaziosmanpaşa Hospital Clinical Researchs Ethics Committee (Number: 995, Date: 09/06/2020).

Informed Consent: Written informed consent was obtained from patients who participated in this study.

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REFERENCES

1. Ma ZL, Yan J, Li SJ, Hua ZD, Yan FX, Wang X, et al. Coarctation of the aorta with aortic arch hypoplasia: midterm outcomes of aortic arch reconstruction with autologous pulmonary artery patch. Chin Med J (Engl) 2017;130:2802-7.
2. Rüffer A, Bechtold C, Purbojo A, Toka O, Glöckler M, Dittrich S, et al. Aortic arch obstruction neonates with biventricular physiology: left-open compared to closed inter-atrial communication during primary repair-a retrospective study. J Cardiothorac Surg [Internet] 2015;10:1-9.
3. Poncelet AJ, Henkens A, Sluysmans T, Moniotte S, de Beco G, Momeni M, et al. Distal aortic arch hypoplasia and coarctation repair: a tailored enlargement technique. World J Pediatr Congenit Heart Surg 2018;9:496-503.
4. Roussin R, Belli E, Lacour-Gayet F, Godart F, Rey C, Bruniaux J, et al. Aortic arch reconstruction with pulmonary autograft patch aortoplasty. J Thorac Cardiovasc Surg 2002;123:443-50.
5. Kostelka M, Walther T, Geerdts I, Rastan A, Jacobs S, Dähnert I, et al. Primary repair for aortic arch obstruction associated with ventricular septal defect. Ann Thorac Surg 2004;78:1989-93.
6. Lai WW, Geva T, Shirali GS, Frommelt PC, Humes RA, Brook MM, et al. Guidelines and standards for performance of a pediatric echocardiogram: a report from the task force of the pediatric council of the American Society of Echocardiography. J Am Soc Echocardiogr 2006;19:1413-30.
7. Backer CL, Mavroudis C. Congenital heart surgery nomenclature and database project: patent ductus arteriosus, coarctation of the aorta, interrupted aortic arch. Ann Thorac Surg 2000;69(4 Suppl):1998-9.
8. Karl TR, Sano S, Brawn W, Mee RBB. Repair of hypoplastic or interrupted aortic arch via sternotomy. J Thorac Cardiovasc Surg 1992;104:688-95.
9. Rao RP, Connolly D, Lamberti JJ, Fripp R, El Said H. Transverse aortic arch to descending aorta ratio: A new echocardiographic parameter for management of coarctation of the aorta in the infant. Congenit Heart Dis 2012;7:160-9.
10. Langley SM, Sunstrom RE, Reed RD, Rekito AJ, Gerrah R. The neonatal hypoplastic aortic arch: Decisions and more decisions. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2013;16(1):43-51.
11. Tulzer A, Mair R, Kreuzer M, Tulzer G. Outcome of aortic arch reconstruction in infants with coarctation: Importance of operative approach. J Thorac Cardiovasc Surg 2016;152:1506-13.

12. Dave H, Rosser B, Reineke K, Nguyen-Minh S, Knirsch W, Pretre R. Aortic arch enlargement and coarctation repair through a left thoracotomy: Significance of ductal perfusion. Eur J Cardiothoracic Surg 2012;41:906-12.
13. Bechtold C, Purbojo A, Schwitulla J, Glöckler M, Toka O, Dittrich S, et al. Aortic arch reconstruction in neonates with biventricular morphology: increased risk for development of recoarctation by use of autologous pericardium. Thorac Cardiovasc Surg 2015;63:373-9.
14. Bromberg BI, Beekman RH, Rocchini AP, Snider AR, Bank ER, Heidelberger K, et al. Aortic aneurysm after patch aortoplasty repair of coarctation: A prospective analysis of prevalence, screening tests and risks. J Am Coll Cardiol 1989;14:734-41.
15. Bernabei M, Margaryan R, Arcieri L, Bianchi G, Pak V, Murzi B. Aortic arch reconstruction in newborns with an autologous pericardial patch: Contemporary results. Interact Cardiovasc Thorac Surg 2013;16:282-5.
16. Kornilov IA, Sinelnikov YS, Soinov IA, Ponomarev DN, Kshanovskaya MS, Krivoshapkina AA, et al. Outcomes after aortic arch reconstruction for infants: Deep hypothermic circulatory arrest versus moderate hypothermia with selective antegrade cerebral perfusion. Eur J Cardiothoracic Surg 2015;48:e45-50.
17. Lodge AJ, Andersen ND, Turek JW. Recent advances in congenital hear surgery: alternative perfusion strategies for infant aortic arch repair. Curr Cardiol Rep 2019;21.
18. Plymale JM, Frommelt PC, Nugent M, Simpson P, Tweddell JS, Shillingford AJ. The infant with aortic arch hypoplasia and small left heart structures: echocardiographic indices of mitral and aortic hypoplasia predicting successful biventricular repair. Pediatr Cardiol 2017;38:1296-304.