Pharmacophore

ISSN-2229-5402

Journal home page: http://www.pharmacophorejournal.com

Pharmacophore
an International Journal
ISSN 2229-5402

A COMPREHENSIVE APPROACH TO THE CORRECTION OF CONGENITAL DEFORMITY OF THE AORTIC ARCH

Fatima Akhmedovna Magomedsalamova[1], Abdulla Magomedovich Kasumov[1*], Khava Magomedovna Kiloeva[2], Akhmad Mukhdanovich Agaev[2], Isa Kazbekovich Abzaev[2], Khadizhat Adlanovna Murdalova[2]

1. ***Department of Therapy, Faculty of Medicine, Dagestan State Medical University, Republic of Dagestan, Makhachkala, Russia***
2. ***Department of Therapy, Faculty of Medicine, Chechen State Medical University, Grozny, Russia***

ARTICLE INFO

Received:
24 Mar 2023
Accepted:
11 Jul 2023

Keywords: Prevention, Congenital deformity, Aortic arch, Aortic kinking, Neurological complications

ABSTRACT

The article is devoted to modern aspects of the prevention of neurological complications during operations for a rare congenital malformation of the cardiovascular system - congenital deformity of the aortic arch, accompanied by compression of the aortic arch, including proximal to the left common carotid artery. The main components of the formation of cerebral circulation disorders at the stage of aortic compression and the role of the autoregulation mechanism are described. The importance of hypothermia, auxiliary artificial blood circulation, and selective perfusion of the branches of the aortic arch in the prevention of disorders of cerebral and spinal circulation was evaluated. The results of the work showed that auxiliary artificial blood circulation allows performing surgical treatment of this complex group of patients with a high degree of protection from ischemic damage to the neurons of the brain. The possibility of selective transfusion of the left common carotid and subclavian arteries allows you to squeeze the aortic arch proximal to these branches and apply anastomoses in convenient conditions without temporary pressure. Careful adherence to the protocol of the operation allows both to significantly reduce the likelihood of severe neurological complications in the early postoperative period and to prevent fatal complications in the long term.

To Cite This Article: Magomedsalamova FA, Kasumov AM, Kiloeva KM, Agaev AM, Abzaev IK, Murdalova KA. A Comprehensive Approach to the Correction of Congenital Deformity of the Aortic Arch. Pharmacophore. 2023; 14(S1): e-723-7281

Introduction

Modern cardiovascular surgery makes it possible to radically cure most congenital and acquired heart and vascular defects, which necessitates a more fundamental study of heart and vascular bed development disorders rarely found in the clinic. However, there are sections in which even surgical clinics with extensive experience significant difficulties. This applies to thoracic aortic aneurysms, especially those involving the arch. Currently, against the background of improved diagnosis of these diseases, there is an increase in the number of surgical interventions for aneurysms [1]. This is explained both by the improvement of the diagnostic base, and the emerging alertness of doctors of many specialties, an increase in the number of degenerative lesions of the aortic wall, including hereditary ones.

Among thoracic aortic aneurysms, a special place is occupied by congenital pathology of the arch and descending aorta. Congenital deformity of the aortic arch (CDAA) is a congenital defect of the aorta, which is characterized by elongation, tortuosity, and kinks of the aortic arch in the pathological structure of its walls. The description of an aneurysm of the distal arch and/or descending thoracic aorta against the background of its numerous bends, and convolutions is found in the works of many foreign and domestic authors [2, 3]. Isolated observations did not allow the authors to draw any conclusions regarding the patterns of development, characteristic clinical manifestations, and the variants of surgical treatment of congenital

Corresponding Author: Abdulla Magomedovich Kasumov; Department of Therapy, Faculty of Medicine, Dagestan State Medical University, Republic of Dagestan, Makhachkala, Russia. E-mail: bucky99@ya.ru.

deformity of the arch and descending aorta. Isolated publications indicate an unfavorable prognosis for patients without surgical treatment in the long term [4].

Being located in the distal part of the aortic arch or the initial part of the descending aorta, CDAA often causes the necessity of clamping the branches of the aortic arch during surgical intervention. This dictates the need for a special attitude to the prevention of neurological complications. High tolerance of the brain to carotid artery compression during ultrasound Dopplerography does not always mean intraoperative safety of the patient, especially at a young age, when tissues are particularly sensitive to ischemia [5, 6]. The work aimed to evaluate the possibilities of various methods of brain protection during operations for CDAA accompanied by compression of extracranial arteries.

Materials and Methods

The work is based on the analysis of 54 clinical observations of patients with CDAA operated in the Dagestan Republic in the period from 2000 to 2020. The average age of patients was 20.8 ± 13.2 years, and more than 70% of patients were under 30 years old. The ratio of women to men was 1:1.

The main instrumental methods of diagnostics were X-ray contrast angiography and, in recent years, computer angiography with contrasting and construction of 3D reconstructions. The distribution of patients by localization was as follows: local aortic lesion between the left carotid and subclavian arteries - 18 people (33.3%), involving the mouth of the subclavian artery and the initial part of the descending aorta - 27 patients (50%), distal to the left subclavian artery – 9 patients (16.7%). Aortic aneurysms developed in the area of the convoluted aorta were noted in 88% of patients, and in patients older than 20 years – in 100% of cases. The average size of aneurysms was 66.4 ± 12.7 mm for the distal part of the aortic arch, and 58.3 ± 9.2 mm for the descending thoracic aorta. Signs of compression effects on the surrounding organs and tissues were noted only in 14 out of 45 patients (31.1%) - 11 patients with aneurysms of the distal part of the aortic arch and 3 patients with an aneurysm of the descending aorta. Important importance was attached to the peculiarities of the brachiocephalic branches. Aberrant movement of the right subclavian artery was noted in 4 patients, in 2 cases there was a departure of the hypoplastic left vertebral artery from the aortic arch, the departure of the left common carotid artery from the brachiocephalic trunk in 1 patient, hypoplasia of the left subclavian artery in 3 patients. Moderate hypoplasia of the descending aorta was also detected in 3 patients.

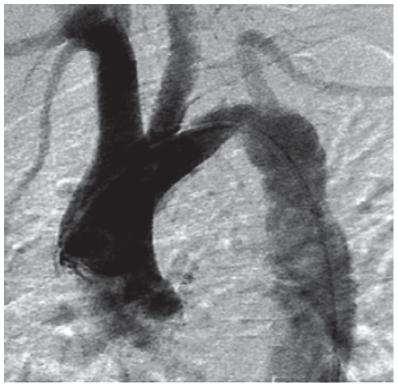
Violation of pulsation of the arteries of the left upper limb (weakening or absence) It was noted in 26 patients, i.e. every second patient has a violation of blood flow in the basin of the left subclavian artery. Violation of blood flow along the right subclavian artery is not characteristic of congenital deformity of the aortic arch. A.lusoria, detected in 4 patients, caused a violation of blood flow through the arteries of the right upper limb only in 2 cases. Thus, 18.5% of patients with CDAA, in addition to the need to pinch the branches of the aortic arch during surgery, had various kinds of abnormalities in the development of brachiocephalic branches.

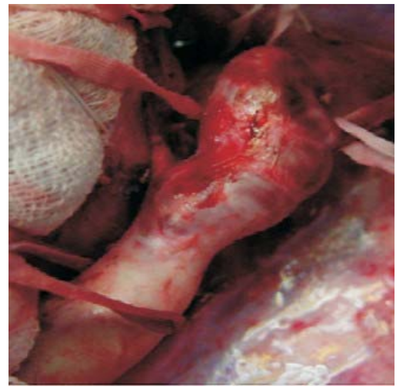
Of the concomitant defects, the following were identified: aortic coarctation - in 10 cases, ventricular septal defect – in 2 patients, tetrad of Fallot, subvalvular aortic stenosis, open oval window in 1 case (27.7%). All patients were operated on. 34 patients were operated on under the conditions of hypothermia, 6 patients underwent normothermia, and 14 patients were operated on using the method of auxiliary artificial circulation (left ventricular bypass and, if necessary, with selective perfusion of the left common carotid and subclavian arteries). Aorta proximal to the left common carotid artery was clamped in 17 patients.

Results and Discussion

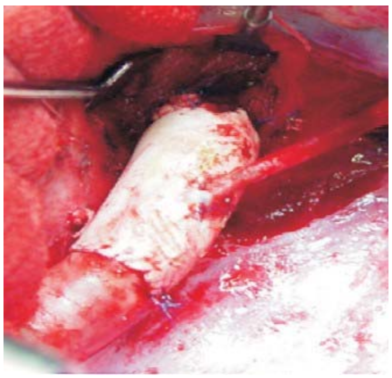
Rare pathology of the cardiovascular system, especially congenital malformations, cause increased attention of specialists in determining the tactics of treatment, and the features of the surgical aid. These aspects, as a rule, are individual and are based on the evaluation of the results of both available research methods and the cumulative experience of the medical institution in the treatment of complex types of pathology of the cardiovascular system. In patients with CDAA, it is very important to assess the location of the branches of the aortic arch in relation to its king, the possible necessity of carotid artery compression, the likelihood of intraoperative complications, and the transition to complete hypothermic circulatory arrest. Planning of brain protection should take into account the factor of arterial hypertension at the stage of aortic compression, the need for compression of the branches of the arch, and the possibility of developing decamping hypotension.

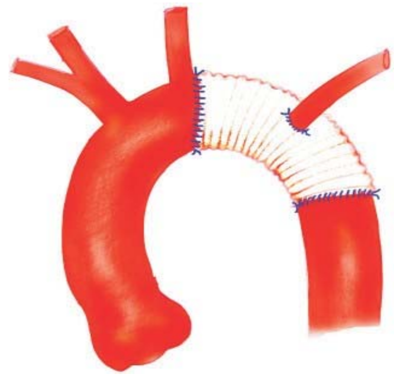
The use of hypothermia in aortic surgery has a long history and is based on a change in the intensity of metabolism in response to a decrease in body temperature. With each degree, the energy demand decreases by 6-7%, and therefore even moderate hypothermia provides an acceptable level of protection for the kidneys and internal organs during aortic compression. Hypothermia also reduces the likelihood of damage to neurons, but it cannot guarantee a sufficient level of metabolism. Thus, from the side of the brain, it is not always possible to determine the minimum safe time level of compression of the branches of the aortic arch. The use of auxiliary artificial blood circulation allows for adequate perfusion of the branches of the aortic arch, preventing the development of ischemia of brain cells. Reconstruction of the distal part of the aortic arch in the case of CDAA of one of the patients is presented in **Figure 1**.





a) b)



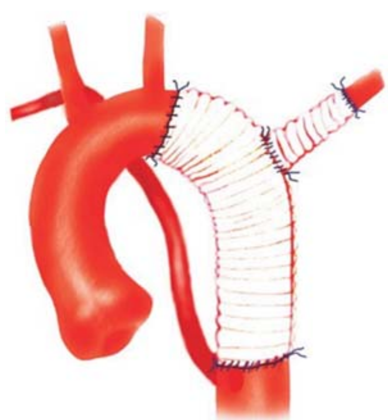


c) d)

Figure 1. Reconstruction of the distal part of the aortic arch in case of CDAA of patient Z.:
a) aortogram, b and c) intraoperative photographs of the stages of the operation, d) the scheme of the intervention

Of 45 patients with localization of congenital aortic deformity in the distal part of the arch, 17 patients required compression of the aorta between the brachiocephalic trunk and the left common carotid artery, i.e. in almost every third case.

The average time of compression of the left common carotid artery was 30.53 = 9.15 min (from 12 to 70 min) for patients operated under hypothermia. In the conditions of normothermia, the carotid artery was squeezed 1 time for 30 minutes without the development of neurological complications (a patient with concomitant aortic coarctation). Acute impairment of cerebral circulation was noted in 2 patients operated under hypothermia, which is 14.3% of 14 cases of compression without perfusion. At the same time, both patients had a negative sample with compression according to ultrasound dopplerography. Intraoperatively, there were no features of the main stage of the operation, the time of compression of the aorta proximal to the carotid artery was 55 and 25 minutes, and the loss during operations was moderate (300 and 500 ml). Aortography data and the scheme of intervention of a patient with an aneurysm of the distal aortic arch against the background of the CDAA and a.luzoria is presented in **Figure 2**.



a) b)

Figure 2. Aortography data (a) and the scheme of intervention (b) of a patient with an aneurysm of the distal aortic arch against the background of the CDAA and a.luzoria

In 3 cases, selective perfusion of the left common carotid and subclavian arteries was used during operations with auxiliary artificial circulation. There were no neurological complications in this group of patients. The absence of a time factor at the stage of applying anastomoses undoubtedly contributes to the quality of their implementation. At the same time, the probability of repeated clamping for applying sealing seams and, consequently, repeated hemodynamic "swings" in the vessels of the brain decreases. Of the specific complications in this group of patients, it is necessary to note the strangulation of the contour in one case due to depressurization with air entering the main line leading to the left common carotid artery. Against the background of the restored blood flow, the air was removed, and perfusion was restored. The stopping time is 3 minutes, there were no neurological complications.

Despite modern advances in medicine in the field of cardiovascular surgery, the risk of developing intraoperative neurological complications during compression of the branches of the aortic arch remains high. The issues of prevention of disorders of cerebral circulation during operations on the thoracic aorta remain relevant at present because this forms not only a significant part of operational mortality but also radically affects the quality of life of those who underwent the intervention. The main and universal damaging agent in the mechanism of formation of cerebral circulatory disorders is hypoxia of brain neurons with subsequent ischemic damage to cell structures. Being a metabolically active organ, the brain receives up to 15% of cardiac output. At the same time, cerebral blood flow in gray matter is 4 times more intense than in white and amounts to 80 ml/100 g/min [7].

The most important point regulating cerebral blood flow is the mechanism of autoregulation, which ensures the constancy of brain perfusion indicators with fluctuations in blood pressure from 50 to 150 mm Hg [8]. The main factors regulating cerebral blood flow are the level of CO_2 and pH in the vessels of the brain. A decrease in the partial pressure of CO_2 by 1 mm Hg reduces the intensity of cerebral blood flow by 1-2 ml /100 g/min, and a drop in $RaSO_2$ from 20 to 40 mm Hg reduces the amount of cerebral blood flow by half. Acute hyperventilation with hypocapnia less than 20 mmHg. it can lead to severe cerebral ischemia caused by vasoconstriction [9].

During the compression of the branches of the aortic arch, there is a sharp, sudden decrease in blood pressure in the main vessels of the brain. The possibilities of autoregulation, in this case, are significantly limited. The time of compression can vary within large limits - from 5-10 minutes to compression over 1 hour. Hypoperfusion is particularly vulnerable to the terminal vascular regions of the brain, where ischemia appears earlier and is more pronounced. At the same time, there is an accumulation of CO_2 and an increase in acidosis, which also reduces the intensity of cerebral blood flow. In addition, the accumulation of under-oxidized products causes edema, which, in turn, can further enhance the violation of perfusion of brain tissues, especially in the cortical layers.

The use of auxiliary artificial blood circulation with selective perfusion of the head branches of the aortic arch makes it possible to level these processes. At the beginning of perfusion, cerebral blood flow even increases due to hemodilution. With the parallel use of hypothermia, the metabolic activity of neurons decreases by about 5-7% with each grade. Autoregulation of cerebral vessels remains constant with changes in perfusion pressure, despite its non-pulsating nature, moderate hyperemia, and hemodilution [10].

One of the main complications of operations using artificial blood circulation is cerebral material embolism. Air, fragments of atherosclerotic plaques, fat bubbles, and microthrombs are an integral part of the use of artificial blood circulation devices. Material embolism is especially common in patients with advanced atherosclerosis and damage to the ascending aorta. In general, any impact on the ascending arch of the aorta can cause an embolism. The use of filters, and membrane oxygenators reduces the number of embolic incidents. In the group of patients with a congenital deformity of the aortic arch, more than 70% of patients were younger than 20 years old. Concomitant ischemic disease on the background of atherosclerotic lesions of the coronary arteries was noted only in 1 patient. Therefore, the risk of material embolism in this category of patients is minimal.

Conclusion

Thus, assisted artificial blood circulation makes it possible to perform surgical treatment of this complex group of patients with a high degree of protection from ischemic damage to the neurons of the brain. The possibility of selective perfusion of the left common carotid and subclavian arteries allows you to squeeze the aortic arch proximal to these branches and apply anastomoses in convenient conditions without temporary pressure. Careful adherence to the protocol of the operation allows both to significantly reduce the likelihood of severe neurological complications in the early postoperative period and to prevent fatal complications in the long term.

Acknowledgments: All the authors were involved in the conceptualization, methodology, formal analysis, writing, and editing of the manuscript.

Conflict of interest: None

Financial support: None

Ethics statement: The processing of operations data was approved by a local Ethics Commission.

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