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**Surgical treatment** **of complex coarctation of aorta with hypoplastic aortic arch in infants**

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 The Article presents the direct experience results of reconstruction of the aortic arch in newborns and infants with coarctation of the aorta (CA) combined with hypoplasia of the arch. In Amosov National Institute of Cardiovascular Surgery were operated 46 children with CA combined with hypoplasia of the arch. For correction hypoplasia of the aortic arch, we have used two methods: in the case of CA and hypoplasia of A and B segments combination, and with the absence of associated congenital heart defects (CHD), we have used left posterolateral thoracotomy according with standard method, which permits to make correction and expand [adequately](http://www.multitran.ru/c/m.exe?t=2858032_1_2&s1=%E0%E4%E5%EA%E2%E0%F2%ED%FB%E9)  hypoplastic segments; in the case of aortic arch hypoplasia completely and with the present of associated congenital heart defects, we have preferred median access using cardiopulmonary bypass (CPB), antegrade cerebral perfusion and one-stage CHD correction.

**Keywords:** aortic arch hypoplasia, antegrade cerebral perfusion, congenital heart disease.

 **Introduction.** According to various authors in 50-81% of cases CA combined with hypoplasia of the aortic arch [1,2,3]. Newborns with this diagnosis is the special children category, which are in critical condition and require urgent surgical intervention. Inoperable newborns mortality average 90% during the first month of life [4]. CA correction without hypoplastic distal aortic arch require reoperation further associated with hypertensive syndrome and an increasing pressure gradient. According to the European database ВВС (ECHSA) 2015, mortality after aortic arch hypoplasia correction with CPB is 17,1 %.

Currently there are several cardiac approaches to solve this problem. Various clinics prefer standard left posterolateral thoracotomy with advanced end-to-end anastomosis, widely used left subclavian artery istmoplastic technique by F. Waldhausen і P. Nahwold [5]. Often the aortic arch and brachiocephalic vessels anatomy prevents to perform adequate mobilization and defects correction by left posterolateral thoracotomy cause a possible risk of unstable hemodynamics in the patient at the time of ascending aorta clamping. In such cases the use of CPB allow safely perform ascending aorta clamping and adequately expand aortic arch by patch or using own tissue of descending aorta for perform expand anastomosis [6]. In addition, concomitant CHD allows them to perform one-stage correction. According to The society of the thoracic surgeons congenital heart surgery database from 2013 amoung 5025 patients isolated CA or aortic arch hypoplasia is found in 54%, combined with ventricular septal defect (VSD) in 17% and combined with another CHD in 29% . It should be noted, that combination of patent ductus arteriosus (PDA), atrial septal defect (ASD), patent foramen ovale (PFO) is apply to isolated CA [7].

According to A. Moulaert et. al., aortic arch hypoplasia exposed when segment C diameter of less than 60%, segment B diameter in less than 50%, segment A diameter less than 40% of the ascending aorta diameter. However, this definition is not quite correct, cause the dilatation of the ascending aorta, which is common in patients with bicuspid aortic valve data are inadequate [8]. Karl et al. offered an original way in which the arch hypoplasia determined by the formula weight in kg+1, measure indicated in mm [9]. The most arch hypoplasia objective criterion is Z score, in which aortic arch hypoplasia is considered -2Z (Z - the standard deviation of the mean).

**Purpose:** present experience of direct results of the aortic arch reconstruction in newborns and infants with CA combined with aortic arch hypoplasia.

**Materials and methods.** In Amosov National Institute of Cardiovascular Surgery were operated 46 children with CA combined with aortic arch hypoplasia from January 2011 until December 2015. The patients' age at the operation time ranged from 1 day to one year, body weight ranged from 2,1 to 10 kg (average weight of 4,8 ± 1,9 kg). Male - 32 children (69%) and female – 14 children (31%). For all patients performed standard echocardiographic examination (echocardiography) before surgery. According to echocardiography, mean pressure gradient at the place of constriction was 55 ± 14.6 Hg.mm. Thus were measured each arch segment, descending and ascending aorta. Besides segmental approach to detect aortic arch hypoplasia, was used calculator Z-score, which takes into account the hypoplasia severity, the degree of stenosis was calculated by the ratio of the aorta segments diameter, deviations from normal performance. Aortic arch segment was considered hypoplastic if Z-score deviation was less than -2.5. Average deviation from normal measures by the Z-score scale: segment A -5,85±1,69, segment B -3,14±1,38, segment C -2,3±1,17. Ejection fraction ranged from 32 to 74%, mean 46 ± 17%. In the study group 65% of patients described pathology combined with the CHD (Tab.1).

Tab.1

|  |  |  |
| --- | --- | --- |
| № | Associated CHD | Amount (%) |
| 1. | VSD | 15 (32,5%) |
| 2. | ASD | 2 (4,3%) |
| 3. | Bicuspid aortic valve | 5 (10,8%) |
| 4. | Transposition of Great Vessels | 3 (6,5%) |
| 5. | Interrupted aortic arch  | 3 (6,5%) |
| 6. | Complete AVSD  | 2 (4,4%) |

In 14 patients additionally performed contrast computed tomography (CT). The data were matched with echocardiography to CT. Results were compared with echocardiography. However, CT results allow more detailed for study of the aortic arch anatomy, select the type of operation, and determine way to CPB connection.

**Surgical management.** Depending on the aortic arch hypoplasia length, CHD concomitant, correction performed by two methods. In the case of CA and hypoplasia of A and B segments combination, and with the absence of associated congenital heart defects (CHD), we have used left posterolateral thoracotomy according with standard method. In the case of aortic arch hypoplasia completely and with the present of associated congenital heart defects, we have preferred median access using cardiopulmonary bypass (CPB), antegrade cerebral perfusion and one-stage CHD correction.

All children operated with endotracheal anesthesia. Anesthesia induction was performed using Sevoflurane (6-8%), maintenance dose - 0.8-1 %. Relaxation was performed using Arduan (dose 0,08-0,1 mg/kg), maintenance dose – 0,04 mg/kg. Analgesia was performed using Fentanil (dose 15-40 mg/kg/h).

In 37 patients for CA with aortic arch hypoplasia correction was performed following method: access - left posterolateral thoracotomy in the third intercostal spaces, was mobilized aortic arch, left subclavian artery, left common carotid artery, descending aorta proximal part. PDA was ligatured and cutted. In necessary, sewed and cutted by 1-2 pairs of intercostal arteries. CA part isolated with careful excision ductal tissues. Aortic arch segments expanding performed on the lesser curvature. Than expanding anastomosis was made by circulate suture. Correction through median access using CPB and antegrade cerebral perfusion was performed in 9 patients. Indications for aortic arch reconstruction through the median sternotomy were: present of combined segments A and B hypoplasia in the patients with CA and associated intracardiac pathology. For all patients was performed right radial artery catheterization before the operation to monitor the adequacy perfusion during aortic arch reconstruction and one of the femoral arteries for direct manometry. Cerebral saturation measured using **Somanetics INVOS.** The sensor emits a beam with a wavelength of 730-810 nm, providing constant regional hemoglobin saturation monitoring in the cerebral cortex. Sensors are placed on both sides of the frontal area, displaying the results on the monitor.

The median sternotomy mobilizes all segments of the aortic arch and brachiocephalic vessels. Ascending aorta cannulation was performed in discharge brachiocephalic trunk area. Cava veins cannulation was performed by the standard method. After CPB start carried out by cooling a patient to 15-18 °. Then arterial cannula was conducted in brachiocephalic trunk and began antegrade cerebral perfusion, reducing the volume perfusion rate to 20- 30% of normal (50-60ml / kg / min). In all cases, after aorta clamping to protect the myocardium using cardioplegic solution by Bretschneider (Custodiol) at the rate of 40 ml / kg. To prevent air embolism during the main phase of the tourniquet clamped aortic cannula and cutted left carotid and subclavian artery. To prevent air embolism during the main phase aortic cannula was clamped by tourniquet and cutted left carotid and subclavian artery. The aortic arch section was performed in the longitudinal direction from descending to ascending region. In 8 patients the aortic arch correction was performed by pericardial patch the previous fixation in 0.6% glutaraldehyde solution. Include to anatomical defects (double aortic arch and hypoplasia of both arcs) in one case held hypoplastic right-arch prosthetics by prosthesis Gore-Tex diameter of 6 mm. After the aortic arch reconstruction and prevent air embolism, arterial cannula transferred from brachiocephalic trunk into the existing aortic arch, thus restoring the estimated systemic perfusion and started warming the patient. At this stage performing associated CHD correction. For all patients was used modified blood ultrafiltration, ultrafiltration volume was 30 to 50 ml / kg.

Tab.2

**Intraoperative measures operated patients with cardiopulmonary bypass (n=9)**

|  |  |
| --- | --- |
| Measure | Mean values (±SD) |
| Operation duration (min) | 223±57 (170-268) |
| CPB duration (min) | 147± 41 (115 - 286) |
| Aortic clamping duration (min) | 81±25 (65 - 128) |
| Antegrade cerebral perfusion duration | 45±17 (32 – 65) |
| Pulmonary ventilation duration | 74,8±18,4 (23-180) |
| The cooling body temperature during the main stage (°C) | 16,5±1,7 (14 - 18) |

 After CPB stopping to assess the immediate results of aortic arch reconstruction measured arterial blood pressure gradient between the radial and femoral arteries. In patients with intracardiac stage reconstruction performed transesophageal echocardiography.

 **Results and discussion.** Hospital mortality is 2,2% (n = 1). The patient was performed aortic arch reconstruction in the median access with the CPB, antegrade cerebral perfusion and narrowing of the pulmonary artery in Taussih-Bing anomalies with aortic arch hypoplasia. The postoperative period was complicated by bilateral pneumonia and sepsis. The patient died on the 75th day caused by growing multiple organ failure.

 Surgical complications were found in 4 (8.6%) patients. In two patients, who has arch hypoplasia correction performed on the left-side access, found chylothorax that was treated by conservative methods (diet, sandostatin). In one patient, whom performed the defects correction in the median access, was found left dome diaphragm paresis and was performed diaphragm plication.

 Respiratory infectious complications in the postoperative period detected in 3 (6.5%) patients: pneumonia -2 patients , catarrhal bronchitis -1 patient.

 Neurological complications from the central nervous system in the early postoperative period wasn’t.

 According infrared spectroscopy sensors results in the brain saturation decrease wasn’t below as 40% during antegrade cerebral perfusion. However, it was observed splanchnic saturation reduction during antegrade cerebral perfusion in 3 patients. At the time of the second blood sampling, before starting the blood flow in the ascending aorta, according to infrared spectroscopy sensors splanchnic saturation measures pair to the normal numbers, in second blood sampling from inferior vena cava venous saturation was high than 40% in all cases.

 Hemodynamically significant pressure gradient in reconstruction region wasn’t found in all patients. During performing echocardiography before discharge pressure gradient in correction region was from 6 to 18 mm Hg (Average 10,5 ± 1,2 mm Hg).

 Including our experience, using sternotomy with CPB and antegrade cerebral perfusion, justified for the category of patients with aortic arch segments hypoplasia and associated CHD. Performed operations through the sternotomy allow to performed arch reconstruction throughout. This access provides good mobilization of all aortic arch segments and brachiocephalic vessels. Using CPB with antegrade cerebral perfusion permit to escape disstable hemodynamics at the time of aortic clamping. In addition, its possible one-stage correction of other heart defects combined with the aortic arch hypoplasia. Traditionally, to protect the internal organs in terms aortic arch clamping was used circulatory arrest combined with deep hypothermia [11]. The main circulatory arrest disadvantages is limited time of cardiac arrest, coagulation hemostasis violation and hypercatecholaminemia [12]. As an alternative approach in 1996 TAsou et al. was proposed the unilateral antegrade selective cerebral perfusion method, which provides continuous blood flow in the brain and saves it in the internal organs and allows to select temperature conditions [11].

**Conclusions:**

1. Using CPB with antegrade cerebral perfusion allow safely and effectively perform the defects correction in patients with the aortic arch segments hypoplasia and associated congenital heart defects.
2. Using antegrade cerebral perfusion combined with deep hypothermia provide adequate brain protection during surgical operation.
3. Defect correction by left posterolateral thoracotomy is indicated for patients with CA and hypoplasia of A and B segments, and with the absence of associated congenital heart defects.
4. The most prefer method of CA verification, CA combined with aortic arch hypoplasia is Z-Score criterion, which is calculated on actual growth and regulatory children body weight.

**Reference:**

1. J. Stark, M de Leval and V.T. Tsang. Surgery for confenital heart defects// Third edition. -2006. –P.285-299.
2. Cruz E. Pediatric and congenital Cardiology, cardiac surgery and intensive care / E. Cruz ., D. Ivy , J. Jaggers .- Springer –Verlag London, 2014. – 3572 p.
3. Wypig D., Nowak C., Colberg C. et al. Extended resection and end-to-end anastomosis for aortic coatctation in infants: results of a tailored surgical approach // Ann Thorac Surg. – 2005. -№80. –P.1453-1459.
4. Constantine Mavroudis. Pediatric cardiac surgery / Constantine Mavroudis, Carl L. Backer // Third edition. -2003. –P. 185-207.
5. Waldhausen F., Nahrwold P. Repair of the coarctation of the aorta with a subclavian flat // A. Thorac. Cardiovasc. Surg. 1966. Vol. 41. P. 425-432.
6. Ascending Sliding Arch Aortoplasty: A Novel Technique for Rep air Arch Hypoplasia / E. D. McKenzie [et al.] // Ann. Thorac. Surg. 2011. Vol. 91 (3). P. 805-810.
7. Ungerleider R., Pasquali S. et al. Contemporary patterns of surgery and outcomes for aortic coarctation: an analysis of the society of thoracic surgeons congenital heart surgery database // T. Thorac Cardiovasc Surg. -2013. – Vol. 1, №145. –P. 1 -20.
8. Moulaert A., Bruins C., Oppenheimer-Dekker A. Anomalies of the aortic arch and ventricular septal defect // Circulation. 1976. Vol. 53. P. 1001-1015.
9. Stark J. Surgery for congenital heart defects / J. Stark, M. Leval, Tsang V.-3rd ed. – John Wiley and sons, 2006. -766p.
10. Smith Maia M., Cortes T., Parga J. et al. Evolutional aspects of children and adolescents with surgically corrected aortic coarctation: clinical, echocardiographic and magnetic reconance image analysis of 113 patients // The J of Thorac and Cardiovasc Surg. -2004. –Vol.127, №3.-P. 712-719.
11. Синельников Ю.С., Корнилов И.А., и др. Защита головного мозга при реконструкции дугы аорты у новородженных // Патология кровообращения и кардиохирургия. -2013. -№3. –С. 4-7.
12. Mossad E., Machado S., Apostolakis J. Bleeding following deep hypothermia and circulatoty arrest in children // Semin Cardiothorac Vasc Anesth. -2007.-№11.-P. 34-36.