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**Results of surgical correction of interrupted aortic arch.**

Borodinova O, Ostras O, Bohuta L, Kurkevich A.

***Ukrainian Children’s Cardiac Center***

The article refers to the results of surgical correction of interrupted aortic arch in 33 patients between 2004 and 2014. Results of the analysis showed no significant difference in survival (71.4% vs 79,0%; p> 0,1) and freedom of unplanned operations (60.0% vs 73,3%; p> 0,1) between the staged and primary complete repair of interrupted aortic arch, and a high percentage of freedom from recurrent aortic arch surgery on regardless the chosen method and strategy of repair (90.0% vs 93,3%; p> 0,1). Taking into account equal safety and different organizational and economic factors a primary complete repair remains an operation of choice.

**Keywords:** *interrupted aortic arch, stage repair, primary complete repair.*

Interrupted aortic arch - is a rare congenital heart defect (CHD) (1% of the all CHD), which is characterized by the lack of continuity between ascending and descending aorta, which was first described by Steidele R.J. in 1778 [1]. This heart defect was classified by Celoria G.C. and Patton R.B. in 1959 into three types: type A (38% of cases), type B (58%) and type C (4%). Interrupted aortic arch can be combined with other complex CHD like aortopulmonary window (APW), an anomaly Taussig-Bing, left ventricle outflow tract obstruction (LVOTO), truncus arteriosus (TrArt) and others. The frequency of Di-George syndrome in these patients is 60% [6].

There are two approaches to the treatment of patients with interrupted aortic arch: primary complete repair and staged repair (interrupted aortic arch repair with pulmonary artery banding and subsequent correction of intracardiac defects). Primary complete repair of heart defects is considered to be the best option by many researchers [2,3]. However, it had been chosen a way of staged repair by 2007 year because of late diagnosis, severe condition of patients at admission due to delayed start of vasoprostan infusion and mechanical ventilation. Surgeons have been performing primary complete repair of interrupted aortic arch since 2007, as a result of prenatal diagnosis and improvement of preoperative management of these patients. Given the different results and conclusions regarding the optimality of one-stage and two-stage surgical treatment of interrupted aortic arch available in modern literature [4,7,8], we analyzed the experience of these patients at the Center.

**Purpose** - to assess and compare the results of staged and primary complete repair of interrupted aortic arch.

**Material and methods.** In the period from January 2004 to December 2014 33 children had surgical repair of the aortic arch in "UCCC". All patients were devided on two groups: group 1 – staged repair (14 patients); group 2 – primary complete repair (19 patients). Structure pathology in operated patients of both groups is shown in Table 1.

Descriptive statistics for continuous data are presented as mean ± standard deviation of the range of values. Assessment of normal distribution was performed by calculating w value in Shapiro-Wilk Normality test. Percentage was used to describe the qualitative features. Comparison groups for qualitative features (survival, freedom from operations, structure pathology) was performed using Fisher's exact test. To assess the reliability of differences between groups for continuous variables (age, weight) at the wrong distribution in both groups was used a Mann-Whitney Utest. For all p criteria were bilateral, the criterion of statistical significance was p <0.05.

Table 1.

Main characteristics of preoperative patient groups.

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
|   | Staged repair(n = 14) | Primary complete repair (n = 19) | All (n = 33) | P-value |  |
| Physiological parameters |  |  |  |  |  |
| Age at the time of surgery, day | 18,8±21,9 (0-157) | 8,4±7,4 (0-38) | 12,8±13,4 (0-51) | > 0,05 |  |
| Weight at the time of surgery, kg | 3,4±0,4 (2,2-2,9) | 3,1±0,4 (1,9-4,5) | 3,2±0,5 (1,9-4,5) | > 0,05 |  |
| Anatomical indicators |  |  |  |  |  |
| Type A | 4 (28%) | 4 (21%) | 8 (24,2%)  |  0,7 |  |
| Type B  | 9 (65%) | 15 (79%) | 24 (72,7%) |  0,7 |  |
| Type C  | 1 (7%) | 0 | 1 (3,0%) | 0,4 |  |
| VSD | 3 (21%) | 3 (15%) | 6 (18%) | 1,0 |  |
| LVOTO | 8 (57%) | 13 (68%) | 16 (48%) | 0,7 |  |
| АPW | 0 | 3 (15%) | 3 (9%) | 0,2 |  |
| Taussig-Bing anomaly | 3 (21%) | 0 | 3 (9%) | 0,09 |  |
| Thymus aplasia  | 6 (42%) | 12 (63%) | 18 (54%) | 0,5 |  |

Correction of interrupted aortic arch and pulmonary artery banding was performed through the left-side thoracotomy, a primary complete repair of the aortic arch and correction all intracardiac defects - through the middle sternotomy. Correction of interrupted aortic arch in the group 1 was performed by anastomosis end-to-side (n = 6), anastomosis end-to-end (n = 3), anastomosis side-to-end (n = 1), extended anastomosis end-to-end (n = 1), the reconstruction of the aortic arch using Gore-Tex prosthesis (n = 3), in group 2 - by anastomosis end-to-side (n = 10), anastomosis side-to-side ( n = 5), reconstruction of the aortic arch using Gore-Tex prosthesis (n = 3), anastomosis side-to-end (n = 1). Pulmonary artery banding was performed in accordance with the formula Trusler G.A. [5].

**Results and discussion.** In the study group the mean follow-up was 4,2 ± 2,4 years: in the group 1 - 6,5 ± 2,5 years, in the group 2 - 3,0 ± 1,7 years. Early mortality in group 1 was 28.6% (4 of 14 patients), in group 2 - 21.1% (4 of 19 patients). Overall early mortality of operated patients was 24.2% (8 of 33 cases). Three patients died from acute cardiovascular failure, three - from competing extracardiac disease (congenital malformations of the digestive tract), one - of infectious complications; one patient died of complex genetic diseases. During the follow-up there were not late deaths. Among 25 surviving patients 8 (32.0%) patients required unplanned reoperation. There were four patients after staged repair who required unplanned reoperation (40.0%) - one on the left bronchus compression, three - on the band compression of pulmonary artery branches. One patient in this group required planned prosthesis replacement of the aortic arch four years after the first operation. There were not unplanned operations after the second stage of correction. There were four (26.7%) patients after primary complete repair who required unplanned reoperations - two due to the left bronchus compression, one - due to necrotic enterocolitis and peritonitis, one – due to the coarctation of the aorta. There were no patient after primary complete repair who had unplanned reoperation on the left ventricle outflow tract obstruction during the follow-up. It should be noted that among patients after staged repair, two (14%) patients were operated without discharge from hospital two days and two months after the first stage due to heart failure; two (14%) patients did not appear for the second stage correction for social reasons [9].

The results of our analysis show no significant difference in survival between the staged repair and primary complete repair (71.4% vs 79,0%; p> 0,1) and there was no significant difference in freedom from unplanned operations between the staged repair and primary complete repair (60.0% against 73,3%; p> 0,1). The results of the aortic arch correction are good regardless of the selected method and strategy of repair: freedom from reoperations on the aortic arch in patients with staged repair and primary complete repair was 90.0% and 93.3%, respectively (p> 0 1).

**Conclusions.**

There was not a significant difference in survival and freedom from unplanned surgery in patients following staged and primary complete repair of interrupted aortic arch.

There was a high percentage of freedom from reoperations on aortic arch in both groups not depending on the method and strategy of correction.

Considering the staged repair always needs additional hospitalization with planned operation, one more incision of the chest and the risk to loose a patient for various reasons between stages of surgery due to the different factors, primary complete repair remains an operation of choice.

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