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INTRODUCTION

Coarctation of the aorta is one of the most common congenital heart defects (1). Its frequency among newborns varies about 20%. This pathology was first described in 1760 by Morgani and more detailed anatomical description was suggested in Paris (2-4). The author suggested calling it – coarctation, “coarctare” from French means narrowing, pressing (5). The first lifetime diagnosis of coarctation of the aorta was made by Legrandin in 1835 (6). A detailed anatomical description was offered by Barie, based on his anatomical study of 86 deceased (7,8). The first successful reconstructive operations were carried out in the middle of the 20th century (in 1945) in the USA and Sweden (9). In the USSR the first resection of aortic coarctation was performed by Gorchitsa V. in 1953. This method included the creation a circular anastomosis (10,11).

After the first operations the success of reconstructive operations became obvious, but it was revealed unsatisfactory long-term results associated with a high frequency of relapses of the disease (12). Further development of the surgical treatment of

Diagnosis of coarctation with hypoplastic aorta arch

Abstract

The tactics of active surgical intervention in the diagnosis of coarctation of the aorta with hypoplasia concomitant arc in the early stages generally leads to significant improvement in long-term results after surgery. This decrease in the number of cases of recoarctation and persistent increase in blood pressure leads to systemic hypertension. Currently there are many techniques that allow to precisely diagnose the stage of hypoplasia of the aortic arch. This allows you to plan the tactics of surgical treatment of the aortic arch in young patients.

Keywords: Aortic arch, hypoplastic aorta, coarctation of the aorta

this pathology was associated with the creation and improvement of new technologies for creating an anastomosis (13).

In modern literature, the role of aorta arch hypoplasia is widely discussed as one of leading risk factors for the development of long-term complications after surgery (14). The combination of coarctation and hypoplasia of the aortic arch is a common finding among the newborns, the prevalence of this combination can occur in 70% of the patients, and this in its turn is associated with a high risk of coarctation development (15-17). The first detailed definition of aortic arch hypoplasia was proposed by Moulaert in 1976. His results were based on the measurement of the outer diameter of specimens at autopsy (18). The anatomy of the aortic arch he divided into three parts, highlighting proximal and distal segments of the transverse part of the arch and the isthmus of the aorta (Figure 1).

According to classification of Moulaert, hypoplasia of transverse part of arc is defined when the outer diameter is 60% or less of the ascending aorta (18).

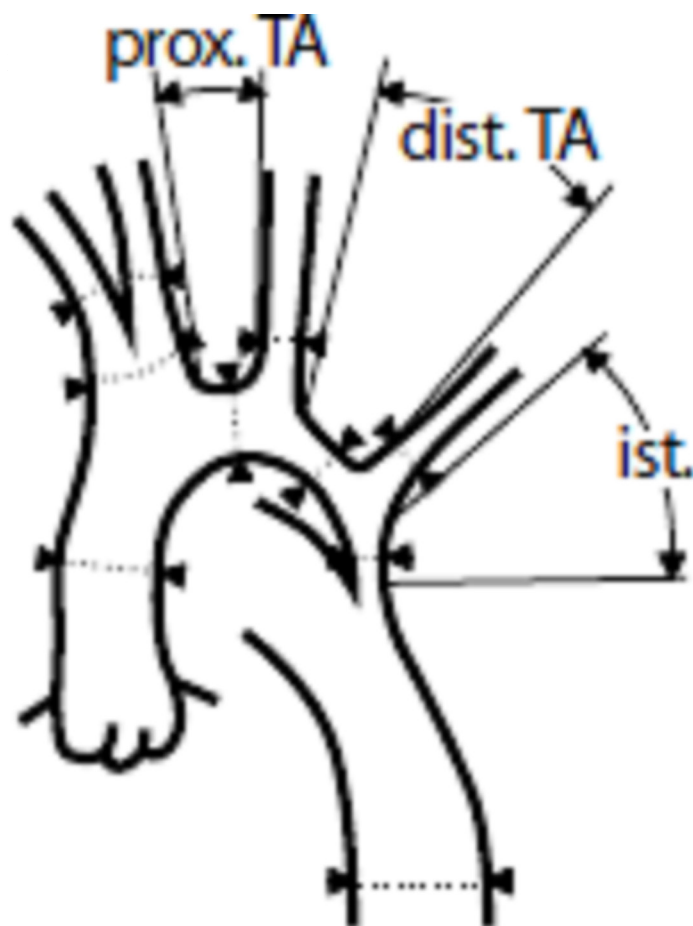


Figure 1. Arch segment of the transverse part: Prox. TA - proximal; dist. TA - distal, ist. – Isthmus of aorta

For distal aortic arch and aortic isthmus, these data should correspond to 50 % or less, 40% and less. In addition, authors of this classification determined the pathogenetic relationship between the presence of a right-to-left shunt and pathology of the aortic arch.

Morphology of the aorta arch hypoplasia

In this study, it was found that in patients with aortic arch hypoplasia there are fewer elastin fibers in its wall, than in comparable segments of the normal arch. The functional significance of this study is significant, since the content of other cellular elements (smooth muscle cells) was also assessed, the density of collagen fibers and the correlation of these elements with the morphological parameters of aortic arch wall segments without hypoplasia were assessed.

In the study by M. Machii et al. It was found that a hypoplastic aorta arch has significantly lower amount of elastin fibers, than a normal aortic arch. At the same time, the size of elastin fibers, in terms of its diameter, exceeds the value obtained in a normally developed aorta. This paradoxical situation described by the authors is specific to tubular hypoplasia of the aortic arch. The phenotypic characteristics of the aortic arch wall cells is

important, since these cells are able to synthesize biologically active substances, thereby supporting proliferation process and activation of the extracellular matrix components, as a result of which they are able to stimulate the further growth process (19).

Pathophysiology

With coarctation of the aorta, blood circulation is divided into two parts (upper and lower) with different blood pressure readings in these parts.

Hemodynamical features of this pathology depend on three factors. These are arterial hypertension of the upper part of the body, a relative decrease of pressure in the arteries of the lower part of the body, and a pressure gradient between the upper and lower circulation. The simplest in term of physiology are cases of mild or moderate obstruction. Systemic hypertension, left ventricular hypertrophy without evidence of acute ventricular failure, and a pressure gradient in the lower and upper extremities result from moderate obstruction.

The pressure gradient in the lower and upper extremities may decrease or even completely level due to the development of collateral blood. The development of collateral blood flow may remain asymptomatic for many years, until the occurrence of complications associated with left ventricular hypertrophy, degenerative changes in the aorta or the development of systemic arterial hypertension (20).

Restriction of blood flow at the level of stenosis creates a pathological systolic pressure gradient between the upper and lower parts of the arterial circulation. Consequently, the cessation of blood circulation occurs in the upper part, results in all parts of the aorta, including the ascending aorta and brachiocephalic vessels, expand compensatory. The arterial vessels of the upper body are in a state of constant hypertension. Blood from the proximal part of the aorta enters its distal part continuously, because the pressure gradient exists not only in systole, but also in diastole. The blood flow creates turbulent eddies in a wide post-coarctation segment, which injures the vascular wall and leads subsequently to the formation of an aortic aneurysm.

This pathogenetic mechanism underlies the formation of aneurysms of the circle of Willis.

Hemodynamic obstruction of the outflow tract of the left ventricle leads to hypertrophy of the left ventricle and the formation of chronic heart failure. Systolic pressure in the ventricle increases according to the level of developed arterial hypertension, the proper ejection fraction is provided by compensatory hypertrophied myocardium. Heart failure develops for a long time, since high diastolic pressure in the ascending aorta provides sufficient coronary blood flow.

Diagnosis of the aortic arch hypoplasia

This disease occurs quite often, this is because during prenatal screening, hypoplasia of the aortic arch is almost impossible to diagnose.

In the present study, the diagnosis was based on the results of 2D Doppler flow echocardiography and MRI. An echocardiogram is also a mandatory procedure, because shows all segments of the aorta, they are obligatory for measurement. Ultrasound examination provides images of the location of aortic coarctation with hypoplasia of the aortic arch and adjacent structures. It may not be possible to visualize this if there is constriction with lung tissue. MRI and MSCT are the standard and most reliable methods for evaluating aortic anatomy (Figure 2). Thanks to these methods, it is possible to determine the localization and length of the constriction, the condition of the branches of the arch, post-stenotic dilatation, and after reconstructive operations, it allows identify aneurysms and recoarctation. Sequential image analysis allows understanding the spatial arrangement of the main structures, their relationship and relation to the mediastinal organs, the bronchopulmonary system.

Initial images in the axial plane do not allow a reliable assessment of this pathology. Therefore, the main data analysis should be carried out on the basis of oblique-sagittal planar MIP reformations built along the axes of the ascending and descending thoracic aorta and three-dimensional reconstructions.

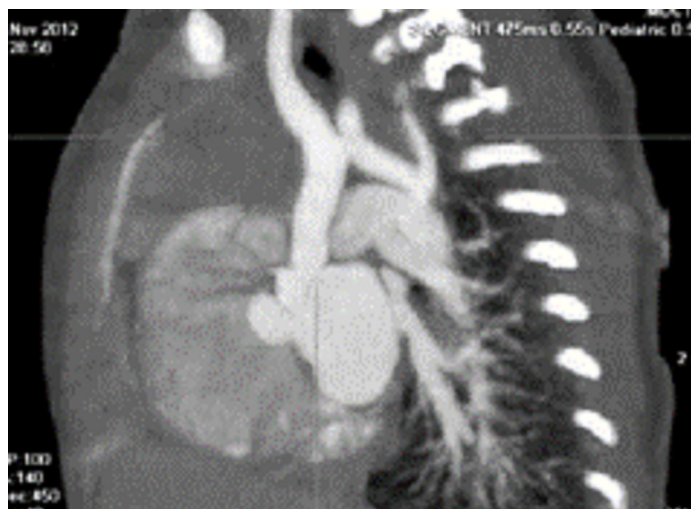


Figure 2. MSCT angiography of the aorta. Hypoplasia of distal segment of the aorta arch

The choice of tactics of reconstructive surgical treatment depends on the degree of hypoplasia and the anatomical features of the arch.

According to the Moulart rule, the diameter of the proximal arch should be greater than 60%, the diameter of the distal arch should be greater than 50%, and the diameter of the isthmus of the arch should be greater than 40% of the diameter of the ascending aorta (Figure 3). If the length of these segments is 5 mm or more, then in this case it is customary to speak of tubular hypoplasia of the arch. According to the above criteria, tubular hypoplasia is defined when this segment is greater than 5 mm and the outer diameter is 40% or less of the diameter of the ascending aorta.

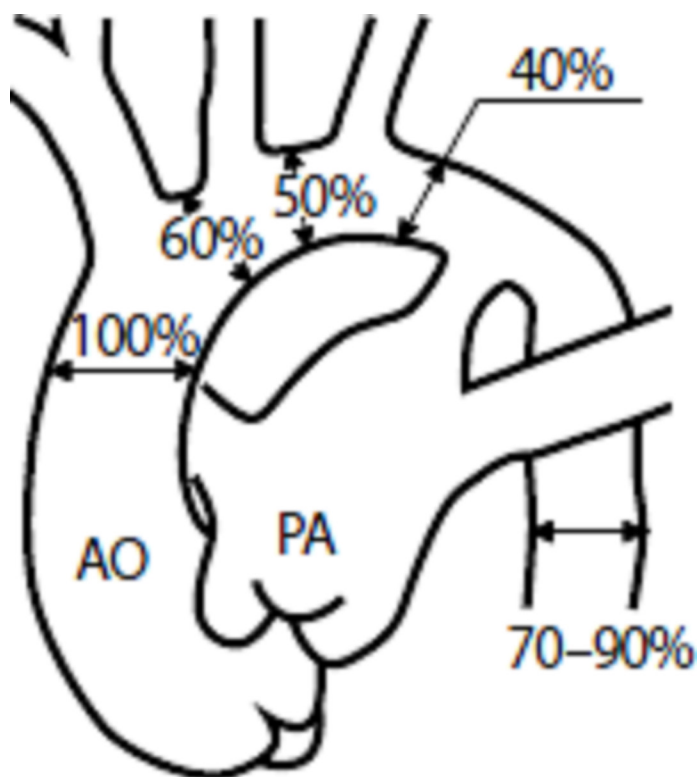


Figure 3. Rule of Moulart

There is another rule that is used in many clinics to determine indications for the correction of arch hypoplasia. This rule was formed by Miyo. Arch reconstruction is necessary if the transverse part of the arch in mm (measured by echocardiography) is less than the patient's weight (kg) plus one. That is, if a newborn weighing 3 kg has an arc diameter less than 4 mm (i.e. $3 + 1$); this is an indication for surgical treatment. Sakurai et al extended this rule by comparing the size of the innominate artery and the size of the transverse fornix. Thus, there is hypoplasia of the arch if the diameter of the transverse fornix is less than the diameter of the innominate artery (mm). In this study, the diameter of the transverse fornix of the arch and isthmus was significantly smaller among patients with coarctation than in the control group ($p \leq 0.001$). In addition, the diameter of the pulmonary valve and its trunk was significantly larger among newborns with coarctation than in the group of healthy newborns. These measurements were standardized by comparing the index (arc index), which is calculated as the diameter of the transverse vault divided by the diameter of the ascending aorta ($\text{INDEX} = \text{TA} / \text{AA}$). This parameter was 0.76 in the control group, and 0.54 in newborns with coarctation. Recently, measurement of the aortic arch and its relationship for the purpose of calculating the exact dimensions of the segment and determining the deviation from the average value of the age norm (Z) are the most popular and accurate.

The aortic arch is considers hypoplastic if the standard deviation $2Z$ or more. It is important to consider that the standard deviation

may change due to body size. It is recommended that Z-scores be calculated in relation to body surface area, rather than separately for height or weight.

Table 1. Results of surgical reconstruction with anastomosis "end to end"

age	patients (n)	Lethality	Recoarctation
<1 m	21	19%	4%
<3 m	12	2%	4%
<6 m	17	1%	9.6%
≤12 m	13	2%	4 %

Table 2. Results of subclavian artery patch repair

age	patients (n)	lethality	recoarctation
<1 m	39	24.8 %	13.6%
<3 m	18	3%	6%
<6 m	16	0	11%
<12 m	11	4%	11%

Table 4. The sensitivity, specificity, positive and negative predictive value of methods

Method for determining hypoplasia	Sensitivity	Specificity	+PC	-PC
Z(score) <-2	97.0%	90.5%	34.2%	99.9%
Morrow hypoplasia index	91.2%	87.6%	29.1%	98.1%
Mee rule	43.2%	94.2%	32.2%	96.4%
Sakurai rule	86.2%	83.7%	22.4%	95.3%

When comparing various methods for determining hypoplasia of the aortic arch with the results of the Moulaert rule, it was found that Z(score)<-2 has the highest sensitivity of 97.0%. The specificity of this technique for calculating hypoplasia was 90.5%, the positive and negative predictive value was 34.2% and 99.9%, respectively.

Thus, to determine the degree of hypoplasia and make a diagnosis of "Hypoplasia of the aortic arch", the Z score can be considered the optimal diagnostic criterion. In our study, we used the Petterson calculator recommended specifically for infants and young children. Calculations are based on Z-value i.e. a value that is defined as the number of standard (signal) deviations from the normal size value for a given body surface area. Z-value < -2 indicates severe hypoplasia.

Modern diagnostic capabilities, medical software, and the introduction of new perfusion support technologies provide a qualitatively new approach to the treatment of patients with

Table 3. Ratio of arch hypoplasia correction from median sternotomy

age	patients (n)	thoracotomy	Sternotomy
<1 m	25	0	100%
<3 m	18	78%	22%
<6 m	16	82%	18%
<12 m	10	76%	24%

The presence of many methods for determining hypoplasia of the aortic arch necessitated the assessment of the prognostic value of generally accepted rules.

To do this, using the example of patients operated on under the age of 1 year, we determined the sensitivity, specificity, positive and negative predictive value of methods for determining hypoplasia of the aortic arch, identified according to the Moulaert rule (Table 4).

Firstly, during the neonatal period, correction of the disease with reconstruction of the hypoplastic arch with cerebral perfusion under cardiopulmonary bypass leads to a significant improvement in the long-term results of surgical correction, the number of cases of recoarctation and the development of systemic arterial hypertension decreases.

aortic coarctation in combination with arch hypoplasia.

Currently, the preference in methods of surgical correction is given to the technique of imposing a long end-to-end anastomosis (Table 1) or applying a patch of the subclavian artery (Table 2) in young children; and for moderate coarctation with arch hypoplasia, balloon angioplasty is the treatment of choice.

When using the patch method, many authors show good results. So 4% mortality and 11% (12/114) recoarctations were reported in the Johns Hopkins Research Group, Baltimore. (21) using this method show the results of zero 30-day mortality and in 11% of cases the development of recoarctation. Our results allow us to say that it is possible to notice a more radical approach through the implementation of sternotomy under cardiopulmonary bypass (Table 3).

Thus, it can be assumed that the correction of coarctation with arch hypoplasia through median sternotomy significantly reduces the risk of recoarctation.

CONCLUSION

This conclusion is consistent with the findings of scientists from the Royal Children's Hospital in Melbourne. Their long-term observations also showed that if the end-to-side approach was performed with a medial sternotomy, the percentage of recoarctations in this group was lower than in the group where the correction was performed via a lateral thoracotomy.

Thus, analyzing our own results and results from various cardiac surgery clinics; we can conclude that the question of the “ideal” method of surgical reconstruction of the aortic arch in case of its hypoplasia among children of the first year of life remains open.

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