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ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY ARISING FROM PULMONARY ARTERY: 10-YEAR EXPERIENCE OF DIAGNOSTICS AND SURGICAL TREATMENT

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The experience of surgical treatment of anomalous origin of left coronary artery arising from pulmonary artery was analyzed in 37 patients. All patients underwent aortic reimplantation of anomalous coronary artery. In early and late postoperative period there was decrease of left ventricular cavity and recovery of left ventricle in most patients. Early diagnostics remains unresolved problem in Ukraine .

Key words: *congenital heart diseases, anomalous origin of left coronary artery, reimplantation of coronary artery*

Anomalous origin of left coronary artery from the pulmonary artery (ALCAPA) is a rare pathology and represents 0,25 – 0,5 % of all congenital heart defects [1]. Not being treated surgically, 90% of patients with ALCAPA die during the first year of life, whereas those who survive the first year suffer from myocardial ischaemia, left ventricular disfunction, mitral valve insufficiency and have a high risk of sudden death[1,6]. Currently the direct aortic reimplantation of anomalous coronary artery (CA) in aorta is the most common surgical technique for ALCAPA in infants and children[2,3,4]. Next to discussable questions regarding surgical correction of this anomaly, there is a problem of early diagnostics of ALCAPA, which is of current interest in Ukraine.

Objective – the objective of this study was to analyze the own experience in diagnostics and surgical treatment of ALCAPA.

Methods and materials. During the period from 2003 till 2013 in UCCC we operated on 37 patients with ALCAPA. Median age of study group was 4 months (9 days to 216 months). Twenty two patients (59,4%) were from 1 to 12 months old. Furthermore in study group there were 13 (35%) patients older than 12 months and 2 (2,4%) were newborns. Median weight was 5.5 kg (2.9 to 69 kg). Among this patients 24 (64,8 %) were females, 13 (35,2%) – males.

To evaluate the ALCAPA diagnostic efficiency we studied patient's medical records and anamnesis by the time of admittance to UCCC.

As a preoperative evaluation methods we used electrocardiography (ECG), echocardiography (ECHO), cardiac catheterization and angiography, computed tomography and magnetic resonance imaging. All patients were surgically treated with direct aortic reimplantation of anomalous coronary artery.

Operative technique. Surgery was done via the midline sternotomy incision. Of paramount importance was putting patient on cardiopulmonary bypass and myocardial protection, especially in patients with ALCAPA and severe left ventricular dysfunction. The key moment was avoiding further myocardial injury. For this purpose during the surgery we were trying to minimize the period between establishing cardiopulmonary bypass and beginning cardioplegia. Optimizing the method, we were putting all the purse-string sutures required and dissecting pulmonary artery branches before establishing cardiopulmonary bypass. Purse-string sutures were put on aorta, inferior and superior vena cava for canulation and also on aorta and pulmonary artery in order to deliver cardioplegia directly in aortic and pulmonary roots simultaneously. After aortic and pulmonary cross-clamping we conducted cardioplegia with the solution cooled to 4°C in a quantity equal to 30 ml/kg. For myocardial protection we used Custodiol, cardioplegic solution of own manufacturing and topical cooling. After the cardioplegia we made full or partial transection of great arteries somewhat above the sinotubular junction. After visualizing the anomalous coronary artery ostium, we conducted its mobilization with sharp dissection and excising its ostium with the part of pulmonary artery wall. When reimplanting CA, to avoid the anastomosis geometry deformation and excessive tissue tension with a risk of postoperative bleeding, we performed the

elongation of the CA in 11 patients, making a tube of pulmonary artery wall, excised with a CA ostium [5]. Like in an arterial switch operation for TGA, we utilized a "trap door" technique in 3 patients with ALCAPA. The pulmonary artery was reconstructed with an autopericardial patch. Surgery was done in a conventional way. We placed a catheter in a left atrium for direct pressure measurement. Delayed sternal closure was performed in 4 patients.

Results and discussion. Diagnostics may be the most challenging issue of the whole curative process. With regard to the low incidence of ALCAPA in a structure of congenital heart defects, only a few specialists in Ukraine have the experience in diagnosing this anomaly, thereof we can face a substantial misdiagnosing during the primary evaluation. When patients were referred to UCCC, ALCAPA was diagnosed only in 7 (18,9%) cases. The most often misdiagnoses were following: dilative cardiomyopathy, diagnosed in 7 (25,9%) patients and myocarditis, diagnosed in 5 (18,5%) patients. Four (14,8%) patients were misdiagnosed with congenital mitral insufficiency, other 4 (14,8%) - with ventricular septal defect, and another 3 (11,1%) – with coarctation of aorta. Among other misdiagnoses were: aorto-pulmonary window, atrial septal defect, aortic stenosis and coronary fistulas. There was no information about the preliminary diagnosis in three patients.

Clinical manifestation was determined by the degree of myocardial ischaemia and congestive heart failure. For the patients under the age of 12 months skin paleness, fatigue, shortness of breath, sweats, loss of appetite, episodes of disturbance due to chest pain and hepatomegaly were common. In elder patients we can see shortness of breath, chest pain, fatigue or they can present with no symptoms at all.

The majority of patients with ALCAPA were admitted with a significant decrease in left ventricular contractility and left ventricular dilation. Median ejection fraction of left ventricle (EFLV) was 33% (від 15 до 64%) median left ventricular end-diastolic index (LVEDI) – 153 ml/ m^2 (15 to 348 ml/m^2). Left ventricular function in patients under the age of 12 months with the infant type of the defect was worse than in older children above 12 months of age with an adult type of the defect (tab.1). Those

differences in groups of different age were due to development of coronary collateral circulation and reduction of myocardial ischaemia with an adult type of the defect [1].

Table 1

Echocardiographic indexes of left ventricle in different age groups

of patients with ALCAPA (before surgery)

Index	age <12 months (n=24)	age >12 months (n=13)
EFLV%	28% (15-47)	51,5%. (20-64)
LVEDI, ml/m2	182 ml/m^2 (15-348)	120 ml/m2(80-312)

In an early postoperative period died three patients. Hospital mortality was 8,1%. In two cases the cause of death was acute left ventricular failure, and in one – pneumonia and sepsis.

Median time of surgery was 270 minutes (180 to 1440 min.), cardiopulmonary bypass duration – 126 minutes (87 to 300 min.), aortic cross-clamping time was – 87 minutes (39 – 116 min.). Median ICU stay duration was 8 days (3 to 44 days), median mechanical ventilation time was 66,5 hours (6 to 720hr.).

In patients with severely decreased left ventricular function during the first 24 – 48 hours we conducted analgesia and deep sedation (morphine hydrochloride, phenthanil, diazepam). In a case of uncomplicated postoperative period, we used light doses of hemodynamic support: dopamine and dobutamine (3-5 mcg/kg/min) and levosimendan (01. – 0.2 mcg/kg/min (since 2006y.)).

At the moment of hospital discharge in 31 (91,2%) patients with ALCAPA there was increase of EFLV, and decreased of LVEDI - in all patients (tab.2). Median EFLV was 50% (21 to 65%), median LVEDI – 147 ml/m^2 (43 to 254ml/m2).

Table 2

Echocardiographic indexes of left ventricle in different age groups

of patients with ALCAPA(after surgery)

Index	age <12 months (n=22)	age >12 months (n=12)
EFLV%	40% (21-65)	59%. (41-67)
LVEDI, ml/m^2	129 ml/m2 (43-256)	80,5ml/m^2(67-185)

Repeat evaluation was made in 28 (82,3%) patients. Median follow-up time was 27 months (3 to 93 months). All patients were in good condition. Median EFLV was 56% (29 to 68%), median LVEDI – 93 ml/m^2 (36 to197 ml/m^2). Compared to preoperative data, the EFLV increase was noticed in all patients, whereas the decrease of LVEDI – in 26 (92,8%) patients.

Conclusion

1. Early diagnostic of ALCAPA remains the unresolved problem in Ukraine.
2. Aortic reimplantanion of CA is effective method of surgical correction of ALCAPA and in most patients it provides ventricular function restoration in postoperative period.

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АНОМАЛЬНЕ ВІДХОДЖЕННЯ ЛІВОЇ КОРОНАРНОЇ АРТЕРІЇ ВІД ЛЕГЕНЕВОЇ АРТЕРІЇ: ДЕСЯТИРІЧНИЙ ДОСВІД ДІАГНОСТИКИ ТА ХІРУРГІЧНОГО ЛІКУВАННЯ

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У статті проаналізовано досвід діагностики та хірургічного лікування аномального відходження лівої коронарної артерії від легеневої артерії (ALCAPA) у 37 хворих. В усіх пацієнтів хірургічна корекція проведена методом реімплантації аномальної коронарної артерії в аорту. В ранньому та віддаленому післяопераційному періоді у більшості хворих спостерігалось

відновлення скоротливої здатності міокарда та зменшення дилатованої порожнини лівого шлуночка. Рання діагностика ALCAPA залишається невирішеною проблемою в Україні.

Ключові слова: *вроджені вади серця, аномальне відходження лівої коронарної артерії, реімплантація лівої коронарної артерії.*

АНОМАЛЬНОЕ ОТХОЖДЕНИЕ ЛЕВОЙ КОРОНАРНОЙ АРТЕРИИ ОТ ЛЁГОЧНОЙ АРТЕРИИ: ДЕСЯТИЛЕТНИЙ ОПЫТ ДИАГНОСТИКИ И ХИРУРГИЧЕСКОГО ЛЕЧЕНИЯ

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В статье проанализирован опыт хирургического лечения аномального отхождения левой коронарной артерии от легочной артерии (ALCAPA) у 37 больных. У всех пациентов хирургическая коррекция произведена методом реимплантации коронарных артерий в аорту. В раннем и отдаленном периоде у большинства больных наблюдалось восстановление сократительной функции миокарда и уменьшение дилатированной полости левого желудочка. Ранняя диагностика ALCAPA остается еще не решенным в Украине вопросом.

Ключевые слова*: врожденные пороки сердца, аномальное отхождение левой коронарной артерии, реимплантация коронарной артерии*