

Kwiatkowska J, Herrador Rey A, Meyer-Szary J, et al. Long-term outcome after surgical repair of anomalous origin of the left coronary artery from the pulmonary artery: 24 years of experience. Kardiol Pol. 2019; Kardiol Pol. 2019; 77: 716-718. doi:10.33963/KP.14816

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Results

The results of the preoperative echocardiography of one patient were unavailable. Left ventricular (LV) dysfunction was present in 17/19 (89%). The mean preoperative LVEF calculated by 2D transthoracic echocardiography (TTE) was 29% (range 13 to 69%). Median LVEDD z-score was 7.66 (range 1.13 to 12.48) and only one patient (1/19, 5%) presented LVEDD z-score lower than 2.0. Preoperative mitral regurgitation (MR) was evaluated by color Doppler flow in TTE and graded as none in 0/19 (0%), trivial in 5/22 (22.7%), moderate in 12/22 (54.5%), and severe in 4/22 (18%) of patients, meaning that 72.7% of patients presented preoperatively with at least moderate MR.

Three out of 22 patients (13.6%) showed associated cardiac anomalies such as an atrial septum defect (ASD) in 1, patent ductus arteriosus (PDA) in 1 patient and multiple ventricular septal defects (VSD) in another one.

The patient with VSD is an example of late manifestation and diagnosis of ALCAPA. The girl underwent banding of pulmonary artery (PA) at the age of 7 weeks and surgical closure of VSD and de-banding of PA at the age of 10 months. The coronary artery anomaly was diagnosed 2 weeks after surgical treatment of the concomitant heart defect.

There were three patients with adult-type of ALCAPA in our study cohort. At the time of the diagnosis of ALCAPA, they were 7, 9 and 13 years old. The first patient, a 7-year-old girl suffered from intermittent angina and irregular heart rate but was in a good clinical condition. The second one, a 9-year-old girl, was consulted by a cardiologist because of chest pain, and in the case of the third one, a 14-year-old girl, the ALCAPA syndrome was discovered during a routine check-up because of a systolic murmur - she was otherwise asymptomatic.

Supplementary Table S1. Baseline patient characteristics

Baseline patient characteristics, n=22	
Female	16 (73%)
Age at diagnosis (months)	5 (3 - 156)
Age at surgery (months)	6 (3 - 167)
Weight (kg)	5,7 (3,1 - 36)
Height (cm)	68 (5,2 - 144)
General status at diagnosis	
severe	8 (40%)
good	8 (40%)
moderate	4 (20%)
Cardiogenic shock at diagnosis	3 (14%)
LVEDD (cm)	4 ± 0,6
LVESD (cm)	3,3 ± 0,7
LVEDD (z-score)	7,1 ± 3,5
LVEF (%)	65 (30 - 83)
LV enlargement	19 (95%)
LV function reduced	
significantly (LVEF??)	16 (80%)
mildly(LVEF??)	2 (10%)
Fibroelastosis	16 (80%)
Aortic stenosis	0 (0%)
Pulmonary stenosis, trivial	1 (5%)
Mitral regurgitation	21 (95%)
trivial	5 (23%)

moderate	12 (55%)
severe	4 (18%)
Tricuspid regurgitation	4 (23%)
trivial	3 (17%)
moderate	1 (6%)
Concomitant congenital heart diseases	3 (15%)
ASD II	1 (5%)
PDA	1 (5%)
VSD	1 (5%)

Supplementary Table S2. Primary cause of hospitalization in a tertiary center

Primary cause of hospitalization in the tertiary centre	n (%)
cardiomegaly on X-ray	7 (35%)
overt heart failure	6 (30%)
dyspnea, heart failure	3 (15%)
heart murmur	2 (10%)
failure to thrive	1 (5%)
extrasystole, supraventricular	1 (5%)
routine follow-up (VSD)	1 (5%)

Supplementary Table S3. Perisurgical details.

Perisurgical details, n=22	n (%)
Surgical procedure	
Takeuchi	4 (18%)
Direct coronary reimplantation	18 (82%)
Early postsurgical complications	14 (64%)
Low cardiac output	12(55%)
Pleural effusion	2(9%)
Perisurgical lenght of stay (days)	25 (8 - 78)
Death	2 (9%)
at 3 months, heart failure and ventricular arrhythmia	1 (5%)
at 21 years, sudden cardiac death (VF)	1 (5%)
One-year follow-up, n=21	
LVEF (%)	65 (30 - 83)

Supplementary Table S4. Patients at last follow-up

	n (%) or median (interquartile)
Patients at last follow-up, n=20	
Female	14 (70%)
Age (years)	14.7 ± 9
Years post surgery	12.5 ± 7.9
Weight (kg)	51.5 (10.7 - 80)
Height (cm)	1.6 (1 - 175)
NYHA II	5 (25%)
Chest pain	2 (10%)
Syncope	2 (10%)
Murmur	9 (60%)
Cardiomegaly by X-ray	3/12 (25%)
Echocardiography*	
LVEDD (cm)	4.4 ± 0.8
LVESD (cm)	2.8 (1.4 – 31.7)
LVEDD (z-score)	0.3 (-1.5 – 4.9)
LVEF (%)	64.7 ± 10.1 (37 - 81)

LVEF reduced	3 (15%)
Fibroelastosis	18 (90%)
Aortic stenosis, mild	20 (10%)
Pulmonary stenosis, mild	6 (30%)
Pulmonary regurgitation	4 (20%)
mild	2 (10%)
severe	2 (10%)
Mitral Regurgitation	15 (75%)
mild	13 (65%)
moderate	2 (10%)
Tricuspid regurgitation, mild	12 (60%)
Baffle leak	1 (5%)

* Echocardiography was performed in a standardized way within the first days post-surgical procedure, and then at 4-6 weeks, 12 weeks, and every 6 months afterwards, or more often if needed.