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**Echocardiographic diagnosis of intramural anomalous origin of the left coronary artery from the pulmonary artery: a single center experience.**

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Background. Anomalous origin of the left coronary artery arising from the pulmonary artery (ALCAPA) is a rare and underdiagnosed congenital cardiac anomaly with potentially severe outcome. Transthoracic Echocardiographic (TTE) diagnosis of ALCAPA is challenging, especially in case of intramural course, often requiring additional imaging techniques.

Aim of the study. This study sought to assess the echocardiographic ability in defining coronary anatomy and intramural course in patients with ALCAPA.

Methods. We retrospectively analyzed records of 13 patients that underwent surgical procedure for ALCAPA at our hospital from May 1999 to March 2015. We excluded from this study one patient due to the association of coarctation of aorta with perimembranous ventricular septal defect. Data on the remaining 12 patients (45 % males, mean age  $66\pm 65$  months, range 0-201 months) was collected and data were retrieved from the clinical records including TTE evaluation, CAT or MRI study, if performed.

Results. Among the 12 patients, eleven (91%) were symptomatic, 1 patient was asymptomatic. At diagnosis, the mean left ventricle ejection fraction was 33.5% (range 10-60%). Mitral regurgitation was present in all patients (mild in 2 (18%), moderate in 7 (64%), severe in 2 (18%)). Three patients (27%) underwent further imaging technique including cardiac CAT (2 cases) and MRI scan (1 case) before surgery. Echocardiographic accuracy of ALCAPA diagnosis was 91,8% as it was surgically confirmed in 11 out of the 12 patients. In four patients ALCAPA was associated with intramural course (ALCAPA-IAR). In 2 of these patients ALCAPA-IAR was suspected at echocardiogram and then confirmed by surgery. In contrast, in 2 patients diagnosis was made at the time of surgery (50% sensitivity and 90 % specificity). Among the 2 patients with echocardiographic diagnosis of ALCAPA-IAR, a CAT scan was also performed in one case before surgery. It is important to underline that correct echocardiographic diagnosis of intramural course were done in the two most recent patients, thus suggesting the existence of a learning curve needed to suspect this very challenging anatomic phenotype.

Conclusion. Transthoracic echocardiography is a reliable tool in detecting ALCAPA and, after a needed learning curve, also in the diagnosis of ALCAPA-IAR.