Diagnosis and/or management of anomalous origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) syndrome in adulthood: A report on twelve cases


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Diagnosis and/or management of anomalous origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) syndrome is extremely rare and no recommendation for its management is available. We undertook this study to analyze initial presentation, management and long-term outcome of adult patients with ALCAPA.

Methods. — We analyzed retrospectively the data of patients hospitalized for a first diagnosis or management of ALCAPA from the charts of cardiology departments of seven French hospitals.

Results. — Twelve adults (30.2 ± 21.4 years) were included. Symptoms consisted of chest pain (58.3%), supra-ventricular arrhythmia (42.9%) and heart failure (33.3%). 60% of the ECG were abnormals. Symptoms consisted of chest pain (58.3%), supra-ventricular arrhythmia (42.9%) and heart failure (33.3%).

Conclusion. — Diagnosis of ALCAPA in adult patients is very rare and is confirmed mainly by coronary angiography in presence of atypical symptoms. Considering its complicated natural course and the good surgical results, surgery should probably be proposed systematically even in asymptomatic adults, in case of accidental discovery.

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Pressure recording analytical method for measuring perioperative cardiac output in pediatric cardiac surgery: A validation study


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Background. — ALCAPA syndrome is a rare congenital heart disease, mainly diagnosed during the first months of life with a high early mortality in symptomatic children in the absence of surgery. Adult form is extremely rare and no recommendation for its management is available. We undertook this study to analyze initial presentation, management and long-term outcome of adult patients with ALCAPA.

Methods. — Observational, prospective and single center study.

Objectives. — Cardiac output (CO) monitoring is important during pediatric congenital cardiopathy surgery. Doppler transesophageal echocardiography (TEE) remains the most valuable technique in perioperative settings. Pressure recording analytical method (PRAM) uses arterial pressure waveform analysis to continuously measure CO. This minimally invasive hemodynamic method was assessed in pediatric intensive care unit but showed controversial results depending on the range of body surface area (BSA). The relevance of the method and the pediatric algorithm remains to be assessed in the perioperative setting.

Methods. — We assessed agreement of PRAM method and TEE in measuring perioperative cardiac output during cardiac congenital surgery in children.

Methods. — Observation, prospective and single center study. Thirty-six children (median [25th—75th percentiles]): sex ratio 14 m/22 f, age 6 years [4 months—15 years]; body surface area (BSA) 0.74 m² [0.50—1.11]) were included. Three to seven measures were simultaneous realized: TEE (aortic diameter followed by transaortic valve continuous wave-Doppler signal via transgastric view) and PRAM pediatric algorithm after sternal closure. Linear correlations, Bland–Altman analysis and percentage of error (Critchley-Critchley) were performed according to BSA (mix and BSA < 1.10 m²).