ALCAPA long term follow-up and prognosis


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The aim of the study was to assess the long term outcome of patients with ALCAPA.

Methods: This study is a retrospective single-center analysis of patients who underwent surgery for ALCAPA from 1980 to 2012. Surgical techniques, demographics, echocardiographic parameters and outcomes were collected. Patients were divided into group I < 2 years at diagnosis, group II : > 2 years).

Results: 48 patients (28 females) were included, median age 6 months (min 4 mos, max 65 y), median weight 6 kg (min 1.9 kg) : 36 in group I and 12 in II. Symptoms of HF were patent in 39 patients (mean age 4 y), 9 were asymptomatic (mean age 10 y) and Q wave on ECG was present in 87% of cases. Mean LVSF = 24.2% (group I = 26.6% vs II = 37.7%, and 16.6% in patients < 6 mos of age), LVEDD and LVESD Z-scores were respectively +2 and +5 in groups I and II; 73% had MR: severe in 6%, moderate in 46%, mild in 21%. Left coronary artery ostium located in the left posterior sinus in 31 cases, right posterior sinus in 12 and in right pulmonary branch in 4. Direct coronary artery reimplantation was performed in 71%, Takeuchi technique in 6%, Meyer technique in 20% and LCA ligation in 3%. Mean age at surgery was 29 mos, mean weight 9 kg. Postoperative mechanical circulatory support was required in 3 cases, who had more severe HF, lower LVSF and longer bypass duration. Mean FU was 81 mos (6 to 312 mos). Freedom from reoperation was 100% at 1 y, 91% at 10 y and 88% at 20 y. LVSF increased by 20% in the early postoperative course and 36% at late FU. MR improved significantly in most of the cases. Overall mortality was 33% (15 in group I died before post-operative Day-30, none in group II), decreasing over time from 55% to 11%, and was lower in patients who underwent direct reimplantation. Q wave disappeared in 82% of the cases; 93% of the survivors were asymptomatic at latest evaluation.

Conclusion: ALCAPA patients have overall good long-term survival and outcome. Age >2 y at diagnosis and direct implantation are factors of favourable prognosis.