Long-term survival and functional status of adult patient with Eisenmenger Syndrome


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In the context of new target therapies, this study aimed to assess the functional status and long-term outcomes of patients with Eisenmenger Syndrome reaching adulthood.

Material and methods: This is a single-centre retrospective review of all patients diagnosed with Eisenmenger Syndrome. Demographics, clinical data, underlying cardiac disease, functional status, therapeutics and outcomes were collected.

Results: 159 patient were included (94 females: 59%), aged 27.7 ± 14.8 years at end-follow up, and 60 with Down syndrome (38%). Underlying cardiac disease was: AVSD in 30%, VSD in 35%, ASD in 9%, PDA in 5%, associated shunts in 5%, complex CHD in 10%, left heart obstruction in 2.5%, pulmonary veins anomaly in 2.5% and TGA in 1%. CHD was native in 122 cases (77%), 7 had palliation (4%) and 30 complete repair (19%). Pulse oxygen saturation was 84 ± 12% (range 44 to 98%), lower in non-operated or palliated cases (81%) than in repaired cases (92%, p = 0.002). Patients were in NYHA class I (18%), class II (42%), class III (37%) or IV (3%), not different with previous repair or not. Target therapy agents were given in 35% of the cases (1 agent in 20%, 2 associated in 13%, intravenous epoprostenol in 1.5%). Death occurred in 26 patients (16%) at the age of 29.3 ± 17.8 years.

Complications occurred in most of the cases (64%) including: hemorrhages events, syncopes, thrombo-embolia, cerebral abscess, infective endocarditis, heart failure or arrhythmias. NYHA class did not differ between patients with or without target therapy. SpO2 was 82% in untreated cases compared to 86% in treated cases (NS). Survival rates were: 98% at 10-years, 93% at 20-years, 87% at 30-years, 83% at 40-years, 73% at 50-years and 53% at 60-years of follow-up. Survival was lower in Down patients (p = 0.0023), in males (p = 0.04) and higher better up to 50-years in patients under target therapy (p = 0.05).

Conclusion: Survival rates of adult patients with Eisenmenger Syndrome seem to improve up to 50-year of age with target therapy agents. These results have to be confirmed by larger scale multicentre studies.