

Long-term outcomes and prognosis of patients with isolated corrected Transposition of the Great Arteries

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This study was to assess the long-term outcome of children and adults diagnosed with congenital corrected TGA (ccTGA) and no significant associated lesion.

Material and Methods: All patients with ccTGA were included in the retrospective single-center study. Clinical features, echocardiographic parameters and long term outcomes were collected. Prognostic factors for poor outcome were assessed.

Results: 55 patients were included (30 males), aged 16.7 ± 18.4 y at diagnosis (0 to 77y, 25% less than 1y of age): 13 (23.6%) diagnosed antenatally, 25 fortuitously (45.5%) and 17 had symptoms (30.9%). Associated mild lesions were present in 23 cases (small VSD, mild pulmonary valve stenosis, small ASD, mild isthmus stenosis) and complete atrioventricular block at diagnosis was present in 7. Follow-up was 17.9 ± 12.7 y. NYHA class I, II, III and IV distribution at onset was respectively: 70%, 18%, 7% and 3.6%, and changed to respectively: 31%, 38%, 24% and 6.7% at end-FU. Echocardiographic RV function was normal on 87% of cases, 13% had moderate and 0% had severe RV dysfunction: percent changed to respectively 73%, 18% and 9% at end-FU. Tricuspid regurgitation was grade 0, 1, 2 or 3 in 33%, 49%, 9% and 9% of cases respectively at onset FU and evolved to 13%, 48%, 24% and 16% at end-FU respectively. HF occurred in 8 cases (14.5%) at the age of 37.5 ± 13.7 y (7 to 61y), AVB occurred in 20 (36.4%) at age 31.4 ± 19.4 y (5 to 72y) and arrhythmias in 16 (29%), over FU. One patient died from uncontrolled HF at 54y of age. Eight patients had surgery (3 tricuspid plasties, 2 tricuspid replacements, 3 PA bandings and 1 heart transplantation). Nineteen pregnancies occurred (11 females) with no death. Echocardiographic RV function correlated with MRI measurements ($r=0.7$, $p=0.005$). RV dysfunction was associated with TR grade ($p=0.025$). HF correlated with TR grade ($p=0.015$) and pacemaker implantation ($p=0.026$). Freedom for HF was 50% at age of 36years.

Conclusion: Isolated ccTGA shows significant worsening of clinical status, RV function and TR grade over twenty years follow-up of this young age population. These data should raise the issue of early LV conditioning for double switch option in isolated ccTGA.